

**8th International
CHARGE Syndrome Conference**

**July 26-29, 2007
Hilton Costa Mesa Hotel
Costa Mesa, CA**



**CONFERENCE
PROGRAM
AND
HANDOUTS**



**The CHARGE Syndrome Foundation, Inc.
www.chargesyndrome.org**

Conference Program and Handouts

TABLE OF CONTENTS

<u>TITLE</u>	<u>PROGRAM SECTION</u>
CONFERENCE PROGRAM	1

<u>Categories of Presentations</u>	
ED=Education or development	MG=Medical or genetic
FS=Family support	NF=New family information
GI=General interest	TA=Transition-to-adulthood

CONFERENCE EVALUATION FORMS	2
GENERAL SESSION KEYNOTE:	

FS “Siblings: Too Important to Ignore” (<i>Tina Prochaska</i>)	3
--	---

<u>ABSTRACTS OF BREAKOUT SESSIONS AND POSTERS</u>	4
--	----------

FRIDAY BREAKOUT SESSION HANDOUTS

-- Friday: Breakout Session #1:

- ED Language Development in CHARGE Syndrome
(*L. Swanson, N. Steele, and W. Keedy*) 5
- MG Clinical and Genetic Evaluation of Children with CHARGE:
the Baylor Study (*S. Lalani*) 6
- GI Life, Liberty, and the Pursuit of Happiness: Is Emotional
Competence Ever an Educational Goal? (*D. Brown*) 7
- FS Grandparent Meeting (*Facilitator: N. Hartshorne*) 8
- FS Fathers Forum (*Facilitator: T. Hartshorne*) 9

-- Friday: Breakout Session #2:

- ED Cracker Barrel Session on Cochlear Implants (*K. Stremel,
N. Hartshorne, E. Taylor, and G. Rodriguez-Gil*) 10
- TA Transition-to-Adulthood:
 - Transition Resources from DB-LINK at the National
Consortium on Deaf-Blindness [NCDB] (*B. McGinnity*)
 - Taking the Fear Out of Sex Education (*Maurice Belote*) 11
- NF CHARGE 101 for New Families: Diagnosis and Anesthesia
(*M. Hefner and K. Blake*) 12

-- Friday, Breakout Session #3:

- ED Strategic Conflict Management Skills in Special Education
Settings (*J. Reiman*) 13
- MG Familial CHARGE Syndrome: Is there a risk of recurrence?
(*C. van Ravenswaaij and J. Bergman*)
- TA Transition-to-Adulthood: Glimmers of Promise
(*J. McNulty and K. McNulty*) 15
- NF CHARGE 102 for New Families: Feeding and Development
(*K. Blake and S. Davenport*) 16

-- Friday, Breakout Session #4:

- MG Panel Discussion on Psychotropic Medicines (*T. Hartshorne, L. Wachtel, and V. Bernstein*) 17
- ED Homeschooling Works: Let the Journey Begin! (*S. Minster*) 18
- TA Transition-to-Adulthood: Getting Connected (*S. Ruzenski, P. Wismer, and C. Kirscher*) 19
- NF CHARGE 103 for New Families: Communication and Transition to School (*R. Last*) 20
- GI Training for Fundraisers (*N. Stanger, S. Perrault, and L. Friedman*) 21

SATURDAY BREAKOUT SESSION HANDOUTS

-- Saturday, Breakout Session #5:

- MG Vestibular Function, Balance, and Development (*L. Travis and J. Thelin*) 22
- ED Strategic Conflict Management Processes in Special Education Settings: IEP Facilitation, Resolution Meetings, and Mediation (*J. Reiman*) 23
- GI The Behavioral Phenotype in CHARGE Syndrome (*T. Hartshorne*) 24

-- Saturday, Breakout Session #6:

- MG Puberty and Growth (including the response to growth hormone) (*J. Kirk*) 25
- ED Panel Discussion on Education and Learning in CHARGE Syndrome (*M. Majors, S. Stelzer, D. Bent, M. Girardi, and E. Breadon*) 26
- TA Creating a Community for Adult Life (*J. Kenley and N. Cornelius*) 27

-- Saturday, Breakout Session #7:

- POSTERS, PROFESSIONALS AND EXHIBITS: Breakout Sessions #7 and #8 (*See ABSTRACTS.*)
- ED Person-Centered Planning (*N. Steele, D. Consacro, and S. Hiscutt*) 28
- MG Pain Issues in CHARGE (*V. Bernstein*) 29

-- Saturday, Breakout Session #8:

- POSTERS, PROFESSIONALS AND EXHIBITS: Breakout Sessions #7 and #8 (*See ABSTRACTS.*)
- MG CHARGE Syndrome: A Neurodevelopmental Perspective: The impact of altered development on learning, behavior, and being (*K. Lauger*) 30
- ED A Wholistic Approach to Developing Educational Plans (*D. Lynne and K. Ziegler*) 31

Conference Program for THURSDAY, July 26th, 2007

2:00 - 6:00	CHARGE Foundation Professional Board Meeting
3:00 - 6:00	Registration
7:00 - 10:00	Reception

Conference Program for FRIDAY, July 27th, 2007

7:00 - 4:00	Registration
7:00 - 9:00	BREAKFAST
7:30 - 8:30	Beach Club registration
8:00 - 11:45	Beach Club Morning Session
9:00 - 9:45	Opening Session (President Neal Stanger)
9:45 - 10:05	General Session Keynote: "Siblings: Too Important to Ignore" (Tina Prochaska)
10:05 - 10:30	BREAK
10:30 - 11:45	Breakout Session #1—See below
11:45 - 1:00	LUNCH
12:15 - 3:30	Sibshop 1 – See below
12:30 - 5:00	Beach Club Afternoon Session
1:00 - 2:15	Breakout Session #2—See below
2:15 - 2:30	BREAK
2:30 - 3:45	Breakout Session #3—See below
3:45 - 4:15	BREAK WITH SNACK
4:15 - 5:30	Breakout Session #4—See below
5:30 -	PARTICIPANTS ON THEIR OWN FOR DINNER
6:00 -	Dinner Meeting of Adults with CHARGE

	Pacific Ballroom I	Pacific Ballroom II	Pacific Ballroom III	Pacific Ballroom IV	Balboa Bay I & II
Breakout Session #1		<i>L. Swanson, N. Steele, and W. Keady</i> Language Development In CHARGE	<i>S. Lalani:</i> Clinical & Genetic Evaluation of Children with CHARGE: the Baylor Study	<i>D. Brown:</i> Is Emotional Competence Ever an Educational Goal?	<i>N.Hartshorne:</i> Grandparent Meeting
Breakout Session #2		<i>K. Stremel, N. Hartshorne, E. Taylor, & G. Rodriguez-Gil:</i> Cracker Barrel Session on Cochlear Implants	Transition-to-Adulthood <i>B. McGinnity:</i> Resources – DB Link <i>M. Belote:</i> Taking the Fear Out of Sex Education in CHARGE	CHARGE 101 for New Families <i>M. Hefner & K. Blake:</i> Diagnosis & Anesthesia	<i>T.Hartshorne:</i> Father's Forum
Breakout Session #3	<i>J. Reiman:</i> Strategic Conflict Management <u>Skills</u> in Special Education Settings	<i>van Ravenswaaij & Bergman:</i> Familial CHARGE syndrome: Is there a recurrence risk? <i>J. Bergman & van Ravenswaaij:</i> Puberty & Smell in CHARGE	Transition-to-Adulthood <i>J. McNulty & K. McNulty:</i> Transition to Adulthood: Glimmers of Promise	CHARGE 102 for New Families <i>K. Blake & S. Davenport:</i> Feeding & Development	12:15 - 3:30 <i>T. Prochaska & C. Robbins:</i> SIBSHOP 1
Breakout Session #4	<i>T. Hartshorne, L. Wachtel, V.Bernstein</i> Discussion on Psychotropic Medicines	<i>S. Minster:</i> Homeschooling Works: Let the Journey Begin!	Transition-to-Adulthood <i>S. Ruzenski, P. Wismer, & C. Kirscher</i> Getting Connected	CHARGE 103 for New Families <i>R. Last:</i> Communication & Transition to School	<i>N. Stanger S. Perrault, & L. Friedman:</i> Training for Fundraisers

Conference Program for SATURDAY , July 28th , 2007

7:00 - 4:00	Registration
7:00 - 9:00	BREAKFAST
8:00 -11:45	Beach Club Morning Session
8:45 -12:00	Sibshop 2 – See below
9:00 - 9:10	General Membership Meeting: Updates and Awards (<i>President Neal Stanger</i>)
9:55 -10:20	General Session Keynote: “Learning from Each Other” (<i>Joe McNulty</i>)
10:20 -10:35	BREAK
10:35 -11:45	Breakout Session #5—See below
11:45 - 1:00	LUNCH
12:30 - 3:45	Sibshop 3 – See below
12:30 - 5:00	Beach Club Afternoon Session
1:00 - 2:00	Breakout Session #6—See below
2:00 - 2:15	BREAK
2:15 - 3:15	Breakout Session #7—See below
3:15 - 3:45	BREAK
3:45 - 4:45	Breakout Session #8—See below
6:00 - 9:30	CARNIVAL, DINNER, & SILENT AUCTION

	Pacific Ballroom I	Pacific Ballroom II	Pacific Ballroom III	Pacific Ballroom IV	Balboa Bay I & II
Breakout Session #5		<i>L. Travis & J. Thelin:</i> Vestibular Function, Balance, and Development	<i>J. Reiman:</i> Strategic Conflict Management Processes in Spec Ed Settings: IEP, Resolution, & Mediation <i>M. Major, S Stelzer, D. Bent, M. Girardi, & E. Bredon</i>	<i>T. Hartshorne:</i> The Behavioral Phenotype in CHARGE	9:00 -12:15 <i>T. Prochaska & C. Robbins:</i> SIBSHOP 2
Breakout Session #6		<i>J. Kirk</i> Puberty & Growth (including response to growth hormone)	<i>N. Steele, D. Consacro, & S. Hiscutt:</i> Person-Centered Planning <i>K. Lauger:</i> CHARGE Syndrome: Neurodevelopmental Perspective	<i>J. Kenley & N. Cornelius:</i> Creating a Community for Adult Life	
Breakout Session #7	POSTERS, PROFESSIONALS, & EXHIBITS			<i>V. Bernstein:</i> Pain Issues in CHARGE	1:00 - 4:15 <i>T. Prochaska & C. Robbins:</i> SIBSHOP 3
Breakout Session #8				<i>D. Lynne & K. Ziegler:</i> A Wholistic Approach to Developing Education Plans	

Conference Program for SUNDAY , July 29th , 2007

7:00 - 9:00	BREAKFAST
8:00 -12:00	Beach Club Morning Session
9:00 - 9:10	General Session (<i>President Neal Stanger</i>)
9:10 -10:10	Special Presentation: “California Dreaming”: Questions Answered by Adults with CHARGE (<i>Facilitators: P. Ryan and M. Hefner</i>)
10:10-10:35	BREAK WITH SNACK AND COFFEE
10:35-11:45	Questions Answered by Professionals (<i>Moderator: J. Thelin</i>)
11:45-12:00	Farewell and 2009 Conference Information (<i>President Neal Stanger</i>)

2007 CHARGE Syndrome Conference EVALUATION FORM

[Complete and return to the Registration Table before you leave or mail to Marion Norbury]

Rating Scale: 5= Excellent to 1= Not good

FRIDAY SESSIONS	Quality of Presentation	Value of Information	Repeat in 2009?
FS Keynote: "Siblings: Too Important to Ignore" (Tina Prochaska)			Yes No
<i>Comment:</i>			
FS SIBSHOP 1 (Tina Prochaska and Carol Robbins)			Yes No
<i>Comment:</i>			
ED Language Development in CHARGE Syndrome (Lori Swanson, Nancy Steele, and Wendy Keedy)			Yes No
<i>Comment:</i>			
MG Clinical and Genetic Evaluation of Children with CHARGE: the Baylor Study (Seema Lalani)			Yes No
<i>Comment:</i>			
GI Life, Liberty, and the Pursuit of Happiness: Is Emotional Competence Ever an Educational Goal? (David Brown)			Yes No
<i>Comment:</i>			
FS Grandparent Meeting (Facilitator: Nancy Hartshorne)			Yes No
<i>Comment:</i>			
FS Fathers Forum (Facilitator: Tim Hartshorne)			Yes No
<i>Comment:</i>			
ED Cracker Barrel Session on Cochlear Implants (Kathleen Stremel, Nancy Hartshorne, Ella Taylor, and Gloria Rodriguez-Gil)			Yes No
<i>Comment:</i>			
TA <u>Transition-to-Adulthood</u>: Transition Resources from DB-LINK at the National Consortium on Deaf-Blindness (Betsy McGinnity)			Yes No
<i>Comment:</i>			
TA <u>Transition-to-Adulthood</u>: Taking the Fear Out of Sex Education (Maurice Belote)			Yes No
<i>Comment:</i>			
NF <u>CHARGE 101 for New Families</u>: Diagnosis and Anesthesia (Meg Hefner and Kim Blake)			Yes No
<i>Comments:</i>			

Rating Scale: 5= Excellent to 1= Not good

FRIDAY SESSIONS (p. 2)

Quality of
Presentation

Value of
Information

Repeat in
2009?

ED Strategic Conflict Management Skills in Special Education Settings (John Reiman)

Comment:

Yes No

MG Familial CHARGE Syndrome: Is there a risk of recurrence? (Conny van Ravenswaaij and Jorieke Bergman)

Comment:

Yes No

MG Puberty and Smell in CHARGE Syndrome (Jorieke Bergman)

Comment:

Yes No

TA Transition-to-Adulthood: Glimmers of Promise (J. McNulty and K. McNulty)

Comment:

Yes No

NF CHARGE 102 for New Families: Feeding and Development (K. Blake and S. Davenport)

Comment:

Yes No

MG Panel Discussion on Psychotropic Medicines (T. Hartshorne, L. Wachtel, and V. Bernstein)

Comment:

Yes No

ED Homeschooling (S. Minster)

Comment:

Yes No

TA Transition-to-Adulthood: Getting Connected (S. Ruzenski, P. Wismer, and C. Kirscher)

Comment:

Yes No

NF CHARGE 103 for New Families: Communication and Transition to School (R. Last)

Comment:

Yes No

G1 Training for Fundraisers (N. Stanger, S. Perrault, and L. Friedman)

Comment:

Yes No

2007 CHARGE Syndrome Conference EVALUATION FORM

[Complete and return to the Registration Table before you leave or mail to Marion Norbury]

Rating Scale: 5= Excellent to 1= Not good

SATURDAY SESSIONS	Quality of Presentation	Value of Information	Repeat in 2009?
TA Keynote: "Learning from Each Other" (Joe McNulty)			Yes No
Comment:			
FS SIBSHOP 2 (Tina Prochaska and Carol Robbins)			Yes No
Comment:			
FS SIBSHOP 3 (Tina Prochaska and Carol Robbins)			Yes No
Comment:			
MG Vestibular Function, Balance, and Development (Lori Travis and Jim Thelin)			Yes No
Comment:			
ED Strategic Conflict Management <u>Processes</u> in Special Education Settings: IEP Facilitation, Resolution Meetings, and Mediation (John Reiman)			Yes No
Comment:			
GI The Behavioral Phenotype in CHARGE Syndrome (Tim Hartshorne)			Yes No
Comment:			
MG Puberty and Growth (including the response to growth hormone) (Jeremy Kirk)			Yes No
Comment:			
ED Panel Discussion on Education and Learning in CHARGE Syndrome (Martha Majors, Sharon Stelzer, Donna Bent, Maryann Girardi, & Elizabeth Breadon)			Yes No
Comment:			
TA Creating a Community for Adult Life (Jackie Kenley and Nancy Cornelius)			Yes No
Comment:			
ED Person-Centered Planning (Nancy Steele, Donna Consacro, and Susan Hiscutt)			Yes No
Comment:			

Rating Scale: 5= Excellent to 1= Not good			
SATURDAY SESSIONS (p. 2)	Quality of Presentation	Value of Information	Repeat in 2009?
MG Pain Issues in CHARGE (<i>Veronika Bernstein</i>)			Yes No
<i>Comment:</i>			
MG CHARGE Syndrome: A Neurodevelopmental Perspective: The impact of altered development on learning, behavior, and being (<i>Kim Lauger</i>)			Yes No
<i>Comment:</i>			
ED A Wholistic Approach to Developing Educational Plans (<i>Donna Lynne and Kim Ziegler</i>)			Yes No
<i>Comment:</i>			
Posters, Professionals, & Exhibits			Yes No

General Comments on the Conference

Location: _____

Hotel: _____

Your Room: _____

Food: _____

Meeting Rooms: _____

Daycare: _____

Information Presented: _____

Activities in General: _____

What would make things better? _____

Do You want to come to the next conference?

Siblings: Too Important to Ignore



Tina Prochaska

Tennessee School for the Deaf

8th International CHARGE Syndrome Conference

Costa Mesa, California

July 27, 2007

Sibling Research

Don Meyer

Unique Opportunities

Unique Concerns

The Longest Lasting
Relationship in a Family...

the Sibling Relationship

Guilt

Feeling responsible for the disability

Survivor's guilt

Guilt over abilities or health

Guilt over typical sibling conflicts

Guilt regarding caregiving

If we support typically
developing sibs as they grow up

We increase the chances
they will elect to remain
lovingly involved as adults

Insight

Appreciation for their siblings' abilities

Appreciation for their family

Appreciation for one's health and
capabilities

Isolation, Loneliness and Loss

Isolation from parents' attention
Isolation from information, the process
Isolation from peers

Planning for the Future

Inclusion in decision-making
Information sharing with young adult siblings
Future child-bearing concerns
Special Needs Trusts

Tolerance and Advocacy

Greater tolerance, compassion and understanding of people
Understanding of the consequences of prejudice
Ardent advocates for people with special needs

Information Needs Vary with Children's Age

Preschoolers
School-age children
Teenagers

Resentment

Loss of parental attention
Unequal treatment and excessive demands
Resentment regarding failure to plan for the future

Avoiding Misconceptions about the Condition

Providing Needed Information to Sibs

Keep the sibling's special needs an open topic

Answer sib's questions about the condition

Provide sibs with written materials

Include sibs in visits with service providers

Determine sib's knowledge of the condition



Opportunities for Sibling Support

Sibling Support Project Goals

To provide brothers and sisters of children with special needs an opportunity to meet other siblings in a relaxed recreational setting

To provide brothers and sisters with an opportunity to discuss common joys and concerns with other siblings

To provide siblings with an opportunity to learn how others handle situations commonly experienced by siblings of children with special needs



Sibling Support Project Goals

To provide siblings with an opportunity to learn more about the implications of their brothers' and sisters' special needs

To provide parents with an opportunity to learn more about common concerns of brothers and sisters



Sibshop Web Information:
www.siblingsupport.org

Tina Prochaska: tinap@tsd.k12.tn.us

University of Washington Study on Impact of Sibshops

90% Said Sibshops had a positive effect on their feelings toward their siblings.

2/3 - Said Sibshops taught them coping strategies.

3/4 - Reported Sibshops affected their adult lives.

94% - Said they would recommend Sibshops to others.





Sibshop Web Information:
www.siblingsupport.org

Tina Prochaska: tinap@tsd.k12.tn.us

PROGRAM SECTION 4

ABSTRACTS OF CONFERENCE PRESENTATIONS

BREAKOUT SESSIONS

FS Conducted by Tina Prochaska and Carol Robbins

SIBSHOP 1 (Friday, July 27 12:15-3:30),

SIBSHOP 2 (Saturday, July 28, 9:00-12:15), and

SIBSHOP 3 (Saturday, July 28, 1:00-4:15)

The sibling relationship is often the longest-lasting (and closest) relationship in a family. Research has shown that siblings of children with special needs have specific concerns that children with typically-developing siblings never face. Siblings of children with special needs are also exposed to unique opportunities that are not available to their peers.

Sibshops are places for siblings of children with special needs to obtain peer support and education in a recreational context. They are not therapy sessions, although many parents report they are therapeutic for their children who attend. Sibshops are led by adult siblings (those who've walked the walk) and are geared toward school-age children. Many times, a Sibshop is the first opportunity siblings have to meet others who have special needs brothers or sisters – those who understand what their life is about. Sibshops give siblings a chance to discuss common joys and concerns as well as learning how to handle situations commonly experienced by siblings of children with special needs.

Sibshops are only for siblings...no parents are invited (sorry!). Sibshops are a chance for the siblings to share “the good and the not so good” aspects of their lives with others who are in their shoes, and have fun doing it!

05 ED Language Development in CHARGE (Lori Swanson, Nancy Steele, & Wendy Keedy)

This presentation will focus on language assessment and intervention techniques for children with CHARGE. First, Dr. Swanson will present the overall findings of a research project conducted at the University of Tennessee. In this study, the language development of eight children (CA: 2-13) with CHARGE was assessed. Specific findings of this study will also be presented in an accompanying poster presentation (Emily King, Lori Swanson, Nancy Steele, & James Thelin) at this conference. Second, Ms. Steele will discuss specific intervention strategies to enhance language development. Specifically, she will describe methods to establish intentionality and to increase the frequency of children's use of symbolic communication. Ms. Steele will focus on “upping the ante” to elicit the most conventional form of communication from the child. Each strategy will be accompanied by a video clip. Finally, Ms. Keedy will present specific conditions (limb apraxia, fine motor control deficits, visual perceptual difficulties, cortical vision impairment, autism) which impede communication development, particularly when the child uses gestures and signs. She will then present methods to overcome these complications such as using hand over hand signing, increased response time, and touch cues.

06 MG Clinical and Genetic Evaluation of Children with CHARGE Syndrome-Baylor Study (Seema R. Lalani)

CHARGE syndrome, reported in 1 in 8500-10,000 individuals, is characterized by the presence of coloboma, choanal atresia, cranial nerve defects, distinctive external and inner ear abnormalities, hearing loss, heart defects, urogenital abnormalities, and growth retardation. We have analyzed clinical information and performed genetic testing of CHD7 gene in 134 children with CHARGE syndrome, enrolled largely through the CHARGE Syndrome Foundation meetings, as well as through physician referrals. This study group includes 7 familial cases, where more than one affected individual was identified. Mutations in CHD7 gene were found in 86/134 (64%) children. Most of the affected children have new mutations in the CHD7 gene, which means that the parents are not affected. We found that almost all children who have three combined features of coloboma, choanal atresia and inner ear abnormalities, have mutations in CHD7 gene. Our results also show that there is no distinct correlation between the severity of the clinical findings and the type of mutations seen in CHD7 gene. Heart defects, coloboma of the eye, tracheoesophageal fistula and facial asymmetry are also commonly seen in children who have mutations in CHD7 gene.

07 GI “Life, liberty, and the pursuit of happiness” Is emotional competence ever an educational goal? (David Brown)

As the world of education becomes ever more fixated on the most superficial, skill-based, and standardized measurable goals, we face the imminent creation of a new outcome-based, teach-to-the-test, “No Child Left Behind” generation, which is unlikely to include many people with CHARGE Syndrome. This session will consider what is often interchangeably referred to as ‘emotional maturity’, ‘emotional intelligence’, or ‘emotional competence’, and will attempt to place this back at the very heart of assessment and teaching where it should always be. There will be an examination of how emotional competence develops, and specifically of what problems frequently arise in this area for infants with CHARGE Syndrome. There will also be consideration of broad strategies that help to counter these problems, including educational strategies, therapy approaches, and support for families and professional teams.

08 FS Grandparents Session (*Facilitator: Nancy Hartshorne*)

Grandparents of children with CHARGE syndrome have a unique experience. Grandparents and other adult extended family members are invited to come to this session to share with one another in a safe, facilitator-guided environment. Come and share, or just come and listen. Either way, you are likely to make a connection of support with others living in your shoes.

09 FS Father's Forum: Fathers in CHARGE (*Facilitator: Tim Hartshorne*)

Does father know best? In the bewildering world of CHARGE it can be hard for anyone to know best. This forum is an opportunity for fathers to share how they do it; how they figure out how to father a child with CHARGE. The session will be facilitated by Tim Hartshorne. Tim is a psychologist and the father of an 18 year old boy with CHARGE.

10 MG Crackerbarrel Session on Cochlear Implants (*Ella Taylor, Kathleen Stremel, Nancy Hartshorne, and Gloria Rodriguez*)

This crackerbarrel will serve as a platform in which families can discuss the following:

- 1. Information that was/was not available to them before they made the decision for implantation,*
- 2. The process that they used to make the decision either for or against implantation,*
- 3. The type of intervention/habilitation that their child received or is receiving,*
- 4. Their child's current outcomes, and*
- 5. Their satisfaction with the process (what works, what didn't work, what they would share with other families considering the process).*

We feel that other families, who may be considering implantation, would greatly benefit from this facilitated discussion. Families may also join a LISTSERV for families to increase their networking. The session was designed to achieve the following outcomes:

- 1. Increase in the knowledge of the children with CHARGE who are implanted,*
- 2. Increase in the knowledge of the different types and intensity of habilitation,*
- 3. Increase in the knowledge of family perspectives.*

11 TA Transition-to-Adulthood:

Transition Resources from DB-LINK at the National Consortium on Deaf-Blindness (*Betsy McGinnity*)

An overview of the information and resources available through DB-LINK

Taking the Fear Out of Sex Education in CHARGE (*Maurice Belote*)

Sexuality is an issue that causes anxiety for families and professionals alike, but families and professionals each have a part in teaching social/sexual education. Families instill in their children the values that are fundamental to becoming responsible, healthy adults. Schools provide information so that students can make informed choices based on these family values. While the presence of CHARGE syndrome raises issues that are unique to each child, we can still plan a comprehensive sexuality program that addresses the need for clearly stated information presented in a non-judgmental way. Sexuality education includes many components beyond what we might remember from our own education. These diverse components can include everything from naming body parts and understanding appropriate touch to abuse prevention and public restroom skills. A comprehensive sexuality program also includes social skills training, since social skills are a critical component in successful living and work experiences as adults. We want children to be safe and to have lives that are free of abuse and exploitation, but we also want to provide the information that our children and students need so that they can choose to have the same wonderful life experiences that any of us would wish for ourselves.

CHARGE 101, 102, and 103 for New Families

The purpose of this series of three workshops is to cover the basics about CHARGE for new families (or review for "older" families). Roughly, the first session (CHARGE 101) will cover diagnosis and features of CHARGE and one important early medical issue, anesthesia. The second session (CHARGE 102) will cover feeding issues and then focus on sensory deficits and how they influence early development. The third session (CHARGE 103) will focus on communication: how do babies communicate and what can you do to facilitate communication. And finally, how to evaluate possible educational placements for your child. Along the way, we will try to answer as many questions as possible.

12 NF CHARGE 101 for New Families

Diagnostic and Other Features (*Meg Hefner*)

A brief history of CHARGE: where did the name come from? How often does it happen? How often does it happen again? Diagnosis: What else can look like CHARGE? What about the new DNA test? What are the features of CHARGE? Which ones are helpful in diagnosis and which ones are most important in management? Once the diagnosis is suspected, what tests should be run? What are the consequences of the features of CHARGE? Bring your questions!!

Anesthesia (*Kim Blake*)

Why is anesthesia important? How many surgeries is my child likely to have? Why is it important to combine procedures with one anesthesia?

14 MG

Familial CHARGE syndrome; is there a recurrence risk? (Conny van Ravenswaaij and Jorieke Bergman)

*Since 2004 we know that CHARGE syndrome is caused by a change in the CHD7 gene. Every one has two CHD7 genes (one maternally and one paternally inherited). A change in **one** of the two CHD7-genes is enough to result in CHARGE syndrome. CHARGE syndrome is highly variable and it is not possible to predict the clinical consequences from the specific change in CHD7.*

Two children with CHARGE syndrome within one family incidentally occur. About 3% of all persons with CHARGE syndrome have a sib or parent who also has this syndrome. We have collected information of eleven such families. This reveals important information on the variability of CHARGE syndrome. Within families all affected family members have the same change in CHD7, while their clinical problems differ a lot. Moreover, we discovered that within families often an unexpectedly mild form of CHARGE syndrome can be found. In such families mildly affected persons were identified through a child with typical CHARGE syndrome. In my presentation I will focus on why CHARGE syndrome sometimes occurs in more than one family member and what the recurrence risk is both in specific situations and in general.

Smell and puberty in children with CHARGE syndrome (Jorieke Bergman and Conny van Ravenswaaij)

Delayed puberty and anosmia (inability to smell) are frequent features of CHARGE syndrome. Puberty and smell disorders are often seen in combination, for example in Kallmann syndrome. We have learned that these seemingly independent features are actually linked (in the brain). In embryogenesis the olfactory system and hypothalamus (the driver of puberty) develop together. CHD7 (the gene involved in CHARGE syndrome) seems to have an effect on this developmental process. In particular, we think that a mutation in CHD7 might lead to disturbed development of the olfactory system and hypothalamus. In children with CHARGE syndrome the hypoplastic olfactory system and hypothalamus give rise to anosmia and delayed puberty. We are investigating whether we can predict spontaneous puberty development with a smell test. In addition I will give background information about olfaction (how do we smell, why is smell important) and puberty (how is puberty initiated and what are the risks of delayed puberty).

15 TA Transition-to-Adulthood: Glimmers of Promise (Joe McNulty and Kathy McNulty)

*Congress is striving to secure better post-school outcomes for children with disabilities through recently passed and pending legislation. These laws include the Individuals with Disabilities Education Act (IDEA), No Child Left Behind (NCLB), the Social Security Act & the Workforce Investment Act (WIA). As the primary advocate for their child, parents need to know how these laws can assist them in obtaining appropriate education and adult services. This session will focus specifically on several new initiatives and key policy changes that offer **Glimmers of Promise** in achieving desired transition outcomes. Time will also be allotted to discuss practical strategies on navigating the journey from education to adult services.*

16 NF CHARGE 102 for new Families

Feeding issues (Kim Blake)

What about feeding? How many kids have feeding problems? What do they do about it? Will my child ever eat by mouth?

Sensory deficits: influences of CHARGE features on early development (Sandra Davenport)

Development: How do all those features of CHARGE influence development? What are sensory deficits? What does DeafBlind mean? What does balance have to do with it? What about weakness? How well can my child do?

17 MG Psychiatric Diagnoses and Psychotropic Medications: A pediatric survey (Lee Wachtel and Time Hartshorne)

Many children diagnosed with CHARGE syndrome demonstrate behavioral difficulties in addition to visual, hearing and other systemic impairments. Previous research has reported that children with CHARGE have increased rates of self-injury and aggression, as well as increased frequency of obsessive compulsive and autism spectrum disorders. This study asked parents to report not only the diagnoses given for their child's behavior problems, but also whether psychotropic medication interventions were prescribed, and which agents were chosen. The results of this study showed that according to parental report, anxiety disorders and pervasive developmental disorders were the most common psychiatric diagnoses assigned, with antidepressant and antipsychotic medications the most frequently prescribed psychopharmacological agents.

18 ED Homeschooling Works: Let the Journey Begin! (Shirley Minster)

Homeschooling is an excellent option for families. Those with children with special needs will discover the positive aspects. Shirley will present the benefits of homeschooling, how to prepare to homeschool, how to choose curriculum, and how to set up a workable schedule. This very practical workshop will help parents who want to begin homeschooling as well as those who are already homeschooling. Helpful hints and real life examples and the question and answer period will be helpful to all.

19 TA Transition-to-Adulthood: Getting Connected (Susan Ruzenski, Philip Wismer, and Cathy Kirscher)

This presentation will provide participants with an overview of the services offered from Helen Keller National Center from both the Center-based and Field based perspective. The Center provides comprehensive vocational rehabilitation services to individuals who are deaf-blind. In addition HKNC plays a vital role as a national collaborator among state and local

providers and consumer organizations in order to optimize services and supports and build local capacity among many communities throughout the United States.

There are a variety of options available to youth during high school summer break. A current student participating in the eight week HKNC Summer evaluation and who attended the two-week teen program last summer will share experiences contributing to his personal vision of success and his goals beyond high school graduation. The value of person-centered planning, peer mentorship and collaborative learning, adaptive technology and community work experience are some of the learning experiences he will be highlighting.

20 NF CHARGE 103 for New Families

Communication, communication, communication; transition to a school setting (Rob Last)

How do babies communicate? How can I help my child communicate? What sort of school settings should we be looking for? How do we evaluate potential school placements? How do kids with CHARGE do over time (Rob's video of the same kids covering 13 years).

21 GI Training for Fundraisers (Neal Stanger, Stephen Perrault, and Lacey Friedman)

The CHARGE Syndrome Foundation, Inc. is heavily committed to fundraising to ensure the we can offer the services – like this conference— that are so valuable to the families and professionals we serve. This session is conducted in response to requests from our members who wish to raise funds for the Foundation. Information will be provided about the Foundation's fundraising efforts and how members can participate.

22 MG Vestibular Function, Balance, and Motor Development in CHARGE Syndrome (Lori Travis & James Thelin)

A substantial number of children with CHARGE syndrome do not learn to walk independently. This may be due to vestibular anomalies, visual deficits, muscular hypotonicity, and/or delayed development. Most children with CHARGE syndrome have not had formal vestibular evaluations that would provide valuable diagnostic information relating to their ability to balance and walk. Vestibular evaluations have not been performed in many cases because the children have been incapable of performing the required tasks. New procedures have been developed that may be suitable for these children, and it may be possible to adapt existing procedures for this special population. It has commonly been wondered if children with CHARGE syndrome experience arrest in their development or if they are severely delayed. Through parent surveys, it was found that children with CHARGE syndrome can achieve gross motor developmental milestones but do so at a slower rate than normal children. A correlation between gross motor development and communication abilities was also found. Recent research is focusing on the vestibulo-ocular reflex in CHARGE syndrome.

23 ED Strategic Conflict Management Processes in Special Education: IEP Facilitation, Resolution Meetings, and Mediation (John Reiman)

A vast array of dispute resolution processes are being developed and implemented across the country that focus on earlier conflict resolution, and that aim to strengthen relationships between parents and school personnel. A continuum of procedures has emerged, ranging from informal preventative processes to more formal hearing procedures. This session will detail the 'what, when, how and who' of three powerful and increasingly used resolution processes (IEP Facilitation, Resolution Meetings, and Mediation) being used across the country to effectively/efficiently resolve conflict in service of meeting the interests and needs of every child.

24 GI The Behavioral Phenotype in CHARGE (Tim Hartshorne)

Individuals with CHARGE frequently engage in challenging behaviors. It is most likely that these behaviors have multiple causes. However, the behaviors are similar enough that they may reflect a "behavioral phenotype", or behaviors that are so characteristic of CHARGE that you could almost diagnose CHARGE on the basis of them. This presentation traces the development of our understanding of behavior in CHARGE, and what we know about the multiple causes of the behavior including issues of attachment, sensory deficits, parenting, stress, pain, illness, communication, self regulation, and cognition. We then propose a seven-part behavioral phenotype. This is a beginning attempt to describe the characteristic behavior that is seen in CHARGE. Individuals with CHARGE are often diagnosed as having autism, ADHD, obsessive compulsive disorder, and tic disorder. The problem with these diagnoses in terms of their usefulness is explored in comparison with the use of a behavioral phenotype. The behavioral phenotype may be used to suggest ways to better prevent and intervene with the challenging behavior when it presents itself. Understanding the behavioral tendencies of individuals with CHARGE can assist parents and educators to develop supportive environments to reduce the incidence of challenging behavior.

25 MG Puberty and Growth in CHARGE Syndrome (including response to growth hormone) (Jeremy Kirk)

Problems with retarded growth (the "R" in CHARGE), and also genital problems (the "G" in CHARGE) are well recognized in the condition, although to date there is little published data. 18 patients (7 male) with CHARGE in the UK have been investigated at 12-20 years of age. Almost all the boys previously had genital problems (undescended testicles &/or micropenis (penis less than 1")). Unlike the boys, where none entered spontaneous puberty, most girls have shown some pubertal development, although usually this has arrested part-way through. Although basal hormone levels tend to be low in both sexes, girls show a greater hormonal (gonadotrophin) response during provocative testing.

This indicates that the pituitary pubertal hormonal drive is reduced more in boys than girls; so-called “hypogonadotropic hypogonadism”. Although some also received growth hormone, many (including those untreated with GH) have achieved heights within the normal range.

Using data from two large growth databases (NCGS & KIGS), 58 patients (31 girls) with CHARGE have been treated with GH. In the US cohort, of 22 tested 17 (78%) were growth hormone (GH)-deficient. Most patients have shown some initial growth response to GH, although the improvement in growth rate is not sustained in all long-term, and final height data is lacking.

26 ED Educational Implications on Learning for a Child with CHARGE (Panel Discussion: Moderator: Martha Majors, Assistant Supervisor Deafblind Program Perkins School for the Blind and Educator/Teacher: Sharon Stelzer, Teacher Deafblind Program Perkins School for the Blind)

Discussion will focus on a variety of teaching strategies that assist children with CHARGE to be more effective learners in the school and home settings. Specific strategies will be defined and examples given as to how to implement these strategies across all learning environments. These examples will be directly linked to the IEP content related to accommodations and specially designed instruction.

The poster session will be more specific and individualized for parents; the educators and parents will have time together to develop IEP content related to accommodations and specially designed instruction. The parent should be able to take home a worksheet that will assist in the development of the IEP.

27 TA Creating Community for Adult Life: Planning an Effective and Exciting Transition from School (Nancy Cornelius and Jackie Kenley)

Moving from school to adult life can be a confusing and overwhelming experience for students with CHARGE Syndrome and their families. Creativity, careful planning, and maximizing local community resources help to ensure positive, exciting outcomes for. By sharing personal experiences and current, factual information presenters will explore: the importance of person-centered futures planning as a tool for developing successful transition plans; what to include in transition plans; identifying individual support needs; and accessing local community resources, and adult service agencies. Students with CHARGE have complex needs requiring interaction and collaboration with professionals from many disciplines and service delivery agencies. It takes thoughtful, comprehensive, long range planning to develop and implement an effectual transition plan. In this session, participants will gain ideas and strategies to assist them in developing exciting and effective transition plans for their young adults with CHARGE as they move from school into adult life.

28 ED Inclusion: Using Person Centered Planning Strategies to Bring Dreams to Reality (Donna Consacro, Susan Hiscutt, & Nancy Steele)

This presentation will showcase effective strategies for building and maintaining a successful inclusive environment for a child with CHARGE Syndrome or other disability. The presentation will include information about the:

- 1. Tools from Person Center Planning (PCP) that have been used successfully to bring family, friends, community members, and school personnel together to plan for one child's future.*
- 2. A family's story of successful inclusion for their child with CHARGE Syndrome.*
- 3. The ongoing process of creating and maintaining a team to support a child's inclusion*

The presenters are collaborative partners who have been members of a team surrounding a child with CHARGE Syndrome who has been fully and successfully included in his age appropriate classroom with typical peers in Sullivan County Schools, Kingsport, Tennessee for 4 years. This presentation will offer information from both the parent and professional perspectives on the importance of Person Centered Planning in building and supporting strong teams for children in inclusive environments.

29 MG Pain Issues in CHARGE (Veronika Bernstein)

A high pain threshold is on the list of the additional features of CHARGE Association, however there is no literature documenting the actual manifestations of pain perception and pain assessment for children with CHARGE. We have multiple anecdotal reports of abdominal colic, headaches and migraines, TMJ (Temporomandibular Joint) pain, discomfort due to constipation, megacolon, reflux, and other conditions.

The effects of pain, especially early pain, are well documented in children. There are many close associations of pain with increased anxiety, hyperactivity, passivity, autistic-like behaviors, depression, and aggressive and self-injurious behaviors.

Dr. Bernstein will present results of a survey of parents and care providers of children with CHARGE related to pain recognition, pain assessment, treatment and management strategies. She will review available pain assessments. An adapted version of pain assessment in children with various communication skills will be distributed to the participants. The participants will be encouraged to create a pain checklist to reflect their child's pain related behaviors and strategies that work and do not work (to use with schools, clinics, and other service providers).

30 MG A neurodevelopmental perspective of CHARGE syndrome: Impacts on learning and behavior (Kim Lauger)

From early in gestation the neurodevelopment of an individual with CHARGE Syndrome is altered. Medical crises and procedures, eating challenges, sensory impairments, balance problems and low tone continue to alter the developmental

course. How do these neurodevelopmental changes alter later learning for language, academics, and social development? What influence do these neurodevelopmental changes have on the behavioral “quirks” seen so often in people with CHARGE? And the big question, what can we do about it? Visual models will be used to answer these questions and to provide a framework for intervention.

Background: This session ties together the behavior and learning challenges seen in CHARGE through the eyes of a parent, a pediatric RN, and an Everyday Scientist and insights gained from the fields of brain and behavioral science, psychology, neurodevelopment, deafblindness, and learning disabilities. The impact of cranial nerve and autonomic nervous system irregularities, and how this involvement can interact with the medical and sensory impairments in CHARGE to result in these common behavioral quirks will be explored.

31 ED A Wholistic Approach to Developing Education Plans (Donna Lynne and Kim Zeigler)

To provide participants with the tools they need in order to pull together an effective team that can:

1. *Identify specific medical and health issues that impact the child's learning;*
2. *Identify specific OT, PT, SLP, Vision and Hearing issues that impact the child's learning;*
3. *Identify specific communication needs that impact the child's learning;*
4. *Identify what works and what doesn't work, thereby*
5. *Developing a wholistic education plan that addresses a child's unique learning needs*

POSTERS

GI CHARGE Syndrome Listserv – Eleven Years of Electronically Uniting Families and Others (Lisa Weir)

This presentation traces the history of the CHARGE Syndrome Listserv, which has been bringing families, professionals, and others with an interest in CHARGE together for the past eleven years. The various aspects of this popular and enduring electronic forum are examined.

ED An Analysis of the Communicative Intentions of Children with CHARGE Syndrome

(Emily King, Lori Swanson, Nancy Steele, and James Thelin)

This project describes a pragmatic analysis of eight children with CHARGE syndrome. The primary purpose of this study was to examine the symbolic and non-symbolic language usage of children with CHARGE and to analyze the child's pragmatic acts (communicative intentions) each time he/she communicated. Two communication samples of each child were collected. Each sample was approximately 30 minutes in length. Each sample was transcribed and coded using Fey's (1986) system of coding assertive and responsive conversational acts. Assertive acts include items such as questions, comments, and disagreements. Responsive acts include responses to questions, etc. All child utterances (spoken words, signs, gestures) were coded. Approximately half of the participants were symbolic, the remaining half non-symbolic. Children who used more symbolic communicative acts, spoken or signed, had a wider variety of communicative intentions and a more balanced usage of assertive and responsive communicative acts. None of the children used all of the communicative intents, and many only used a few of them.

There are three possible reasons why children with CHARGE do not frequently use all of the different types of intentions: 1. They do not have symbolic language, 2. They do have symbolic language but have a limited vocabulary or limited use of grammatical forms, or 3. They have limited opportunities to use these communicative intentions. Observations of effective communication strategies to promote symbolic communication are also described.

ED Vision Impairment in CHARGE Syndrome (Donna Bent)

The influence of vision impairment, from mild to severe, in children with CHARGE is significant. Although the multiple issues are difficult to isolate as the entire learning process is impacted, the following stand out as critical factors in the education of this child: movement through space, orientation to the environment, safety, learning media, organization of visual information, sensory issues surrounding eyeglasses and visual input, and overall stamina. This poster will provide the opportunity to see how a comprehensive program of clinical low vision personnel and education specialists at the Perkins School address these issues. The poster session will include functional vision assessment techniques, findings from recent vision acuity research with Db children, Orientation and Mobility curriculum ideas.

MG Puberty in CHARGE syndrome (Jeremy Kirk) See abstract for Breakout Session 25.

FS The experience of siblings of children with CHARGE (Tracy Olson and Tim Hartshorne)

The purpose of this research was to develop a better understanding of how growing up with a sibling with CHARGE syndrome affects a person. This pilot study of 14 siblings examines a number of variables including career choice, loneliness, network orientation, family stress, and coping.

ED Continued Development in CHARGE Syndrome (*Nancy Hartshorne and Joshua K. Hartshorne*)

The results of a longitudinal study of what were originally 100 children with CHARGE syndrome ranging in age from 3 to 15 years will be presented. This is the third in a group of time series studies looking at adaptive behavior and overall development in individuals with CHARGE syndrome. Correlates of developmental abilities in this population will be highlighted, as well as new information gathered about reading abilities, school settings attended, friendships, and adult housing choices.

GI CHARGE Syndrome Listserv – Eleven Years of Electronically Uniting Families and Others (*Lisa Weir*)

MG CHD7 binds to specific sites on chromatin (*Peter C. Scacheri*)

In 2004 it was shown that de novo mutations in the CHD7 gene cause most cases of CHARGE syndrome, but little information has been available about the normal function of the protein. Based on homology to other proteins in the CHD family, we hypothesized that CHD7 is located in the nucleus and is associated with chromatin. To address these hypotheses we made antibodies to CHD7 and tested them by subcellular fractionation and Western blot analysis as well as by chromatin immunoprecipitation on tiled microarrays (ChIP-chip). Our results from mouse ES cells indicate that CHD7 is a nuclear protein that is frequently associated with chromatin at transcriptionally active gene promoters. Some of the CHD7 target genes suggest clues to mechanisms underlying the complex phenotype of CHARGE syndrome. We hypothesize that CHD7 is involved in transcription regulation, and that haploinsufficiency of CHD7 affects expression of multiple CHD7 target genes during development, leading to birth defects in several organ systems.

EXHIBITS & DEMONSTRATIONS

CHARGE Syndrome Revisited: An Instructional CD ROM (Dr. Jan van Dijk, Dr Catherine Nelson, and Arno de Kort MD)

In 2002 the authors produced for the US Charge Conference a cd rom which dealt mainly with challenging behaviors, in particular self-abuse. In the 2005 version the content has been broadened and topics such as literacy and cochlear implants were included as well. In the newest edition (July 2007) 4 of the original children have been recorded again and 2 new children were added. In relation to the latter group important issues such as attachment to the principal caregiver (mother) are discussed.

On this interactive cd rom the authors emphasize the effect of intervention strategies, relating to Challenging behaviors, feeding problems, depression, Cochlear Implantation, development of literacy, and communication. The first part of the cd rom deals with the medical (genetic) background of Charge Syndrome in a clear and understandable way. This cd rom is useful for parents and other day-to-day educators, because it provides very practical information on the upbringing of a child with CS. Many relevant publications are directly linked with the internet.



PROGRAM SECTION 5

Breakout Session #1
Friday, July 27, 2007, 10:30-11:45
Pacific Ballroom II

Language Development
in CHARGE



Presented by
Lori Swanson,
Nancy Steele,
and Wendy Keedy

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Language Development in Children with CHARGE Syndrome

Lori Swanson, Nancy Steele, & Wendy Keedy
8th International CHARGE Syndrome Conference
Costa Mesa, CA
July 27-29, 2007

Acknowledgments

- James Thelin
- Emily King
- Beth Schulte & Page Oliver
- Kentucky Deaf-Blind Project, Tennessee Deaf-Blind Project, South Carolina Deaf-Blind Project
- Parents and Children who participated

Purpose

- Primary purpose was to describe the communication skills of eight children with CHARGE syndrome using the analysis of communication samples as well as parental report.
- Secondary purpose was to begin establishing a valid means of assessing the communication skills of children with CHARGE.

Participants

- 8 Children with CHARGE (CA: 1:8 – 13:1)
- 3 females, 5 males
- All families participated in the Kentucky CHARGE weekend

Method

- Parental questionnaires were completed.
- Two communication samples were collected for each participant.
- Each sample was ½ hour.
- Each sample was transcribed orthographically.
- Each sample was analyzed using Systematic Analysis of Language Transcripts (SALT).

Results - Communication Skills

- 7 of 8 children considered intentional, the remaining child used occasional intentional attempts
- Half of participants considered symbolic, the remaining half non-symbolic

Mode of Communication

- **Symbolic**
- P4 – Primarily signs, rare vocalizations, gestures, noncompliant behaviors
- P6 – Primarily signs, occasional faint vocalizations, head nods/shakes
- P7 – Half gestures, half signs, some vocalizations, some crying
- P8 – Primarily signs, very rare vocalizations, uses facial expressions!
- **Minimally Symbolic**
- P2 – Primarily gestures, some signs, noncompliant behaviors, some self-injurious behaviors
- P3 – Gestures, noncompliant behaviors, self-injurious behaviors
- **Non-symbolic**
- P1 – Gestures, noncompliant behaviors, self-injurious behaviors
- P5 – Gestures, crying, screaming

Summary of Language

- 7 intentional, 1 occasionally intentional
- 4 symbolic, 2 minimally symbolic, 2 non-symbolic
- Primary Mode: 0 speech, 4 sign, 1 gestures, 3 gestures/inappropriate behaviors

Intentional Attempts

- Mean number of intentional behaviors/minute – 2.15 per minute
- Range of intentional attempts per minute (gestural, signs, and verbal) - .70 – 4.5 per minute

Discussion

- Wide range of communication abilities observed
- Symbolic skills appeared related to walking, aural habilitation, early intensive language intervention (consistent with Thelin and colleagues' earlier work)
- For their age, the 2 youngest were most successful – probably due to early intervention.

Discussion

- Limited repertoire of communicative functions, particularly for minimally and non-symbolic children.

Discussion

- Unfamiliar interactant (Nancy Steele) was more successful than classroom teachers at eliciting intentional communication acts from children who rarely initiated.

LANGUAGE DEVELOPMENT IN CHILDREN WITH CHARGE SYNDROME...

*Wendy Keedy
July 2007 Costa Mesa, California*

When searching for hidden treasure
it is often necessary to ask for
assistance...

Perhaps from a "detective". It is perhaps, just
so, as we search for ways to encourage a
child's development of language. It's
important to sift through details and
observations from the parents, teachers and
other providers, plus the child himself.

2

TODAY'S OUTLINE

- ❖ Discuss factors that may limit sign acquisition
- ❖ Describe and demonstrate communication protocols that facilitate communication and improve sign acquisition

3

Some factors affecting sign learning and
who typically identifies the condition

- ❖ Limb apraxia (Neurologist, OT)
- ❖ Fine motor control (OT)
- ❖ Visual perceptual difficulties (Optometrist, Neurologist)
- ❖ Cortical Vision Impairment/Neurological Vision Impairment (NVI) (Ophthalmologist/Neurologist)

4

More factors affecting sign learning

- ❖ Vision loss (Ophthalmologist or optometrist)
- ❖ Receptive Language Disorder (SLP)
- ❖ Expressive Language Disorder (SLP)
- ❖ Autism (Psychologist, Doctor with input from SLP)

5

What is Limb (Idiomotor) Apraxia?

"Praxia" refers to planning. Limb apraxia involves "Motor planning deficits" of the limbs. There is no loss of the ability to perform an action automatically, but the action cannot be performed on request. Unaddressed in a child learning to sign might lead to problems with social interactions and language development.

6

Fine Motor Control

❖ Define

- ❖ Fine motor control may affect creation of signs. Problems may affect forming manual alphabet or individual signs.

7

Visual-Perceptual Difficulties

Vivienne Ratner, Ph.D. in 1988 and 1983 describes a condition she found in 17 % of 3,000 deaf children as: Visual Perceptual Learning Disabilities. These disabilities seriously affect the child's ability to communicate in sign language. The effects can be devastating as the ability to communicate and understand a message that is manually signed is disrupted.

8

"These kinds of difficulties in hearing children have serious ramifications and the ramifications are even more devastating in a deaf child."

from Understanding Language Disorders by Ratner and Harris, 1994

9

"Children with visual perceptual deficits may have incomplete knowledge of their own bodies, resulting in an inaccurate awareness of body parts, their boundaries and their left and right sides.

Consequently these children have difficulty learning to use sign and read messages conveyed in sign by others."

Vivienne Ratner

10

In Understanding Language Disorders, Ratner states

"The children may not discriminate the fine changes in size, shape, or movement of the facial features of a person who is signing, features that are essential to meaning in American Sign Language..."

Ratner found that "45 % of the deaf students in her studies identified as having these forms of visual perceptual disabilities did not learn or comprehend sign language."

11

And that....

"Speech-reading ability is strongly affected by a visual perceptual disability. Children who have difficulty distinguishing between shapes such as a circle and oval, or a square and a rectangle (which are stationary), will certainly have difficulty distinguishing moving and changing oral configurations when speech reading."

12

Effects on IQ of Children who are Deaf with Visual Perceptual Disabilities

Consider that discrepancies between child's level of intellectual function and academic achievement is a traditional way to identify a learning disability. However, in performance tests used with deaf children the tasks require CONSIDERABLE spatial and other visual perceptual skill, such as the ability to discriminate shapes, forms, designs, and to match patterns. These are impossible tasks for an individual with shape discrimination deficits.

13

Effects on Test Scores

The performance scores may then be reduced by the difficulty and the academic performance may be poor so the difference between the two may not reflect accurately a learning disability. The problem may be instead attributed to cognitive disabilities.

14

Visual Spatial Difficulties

"This inaccuracy may result in inappropriate class placement or labeling that can ultimately cause irreparable damage to the child. Using performance scales for deaf individuals renders the scores invalid *unless* visual perceptual learning disabilities have been ruled out."

15

Neurological Vision Impairment (Cortical Vision Impairment)

- ❖ Sam Morgan, Educational Specialist, writes that children with NVI comprise the major cause of vision impairment in children who are deafblind. It is also the major cause of vision loss in children in general.
- ❖ Etiologies can include: lack of oxygen before, during or after birth, viral or bacterial illness such as meningitis or cytomegalovirus or traumatic brain injury.

16

NVI

- NVI effects vision in many ways and can be mild to severe. It may be temporary or permanent. Children with NVI may be more able to see objects peripherally.
- Children may have difficulties with certain types of tasks including figure-ground, complex visual displays such as cluttered pictures and may have spatial confusion.

17

NVI

According to Dr. Christine Roman, children with this condition often look away when reaching for something. She believes that significant changes can happen in brain processing ... especially before age 3. But she doesn't rule out the possibility of changes later on.

18

Autism or Autism-Like Conditions

- ❖ Children might have varying degrees of difficulty with language and with social interactions. They will typically have stimming types of behaviors.
- ❖ Difficulties with vision or visual perceptual difficulty might be associated with autism.

19

Approaches to Consider in Facilitating Communication Growth Limb Apraxia

- Δ Always positive - provide encouraging support
- Δ Hand over/under hand therapy
- Δ Carrier Phrases Δ Prompts Δ Mirror use
- Δ Repetition of words/phrases child signs with confidence Δ Practice predictable phrases from books, rhymes or songs

20

Approaches to Consider in Facilitating Communication Growth Fine Motor Skills

- ❖ Pre-Sign Language Motor Skills, by Marsha Dunn Klein: identifying motor strengths, weaknesses relative to signing. Describes difficulties of 1 hand vs 2 handed signs, motions and positions - written from perspective of occupational therapist.
- ❖ Consider adaptive signs. Match hand to model.

21

Approaches to Consider in Facilitating Communication Growth Visual Perceptual Difficulties

- ❖ Hand over hand and hand under hand
- ❖ Address vestibular issues. Consider that the child may have better access to information and more success signing while sitting rather than standing.
- ❖ Present material in various modalities.

22

Approaches to Consider in Facilitating Communication Growth NVI

- ❖ Materials should be simple in form
- ❖ Bright lighting can help child see and attend to visual materials. Consider natural lighting
- ❖ GIVE THE CHILD TIME TO RESPOND to the materials (Latency)
- ❖ Color vision is usually intact and color can be used effectively
- ❖ Touch should be considered as a *major* sense for learning.

23

Approaches to Consider in Facilitating Communication Growth More NVI

- ❖ Repetition and routines can help child understand visual environment. Repetition can increase comfort level and decrease tension or stress.
- ❖ Try to reduce fatigue by working in short spurts or divide a long task into shorter periods.
- ❖ ...Objects can be more easily seen when moving especially in peripheral fields.
- ❖ Positioning/posture is important. The more energy used to hold oneself up, the less can be used for seeing...

24

Approaches to Consider
in Facilitating Communication Growth
NVI

- ❖ Vision protocols might be suggested by vision specialist. To increase pertinence, remember to make them happen in everyday, real-life situations, not only in therapy sessions. Some examples are: visually tracking classmates as they move across a room, and identifying, signing, the shape of everyday objects.

25

Approaches to Consider
in Facilitating Communication Growth
Autism or Autism-like conditions

- ❖ It is vitally important to identify ways in which the child responds to language. Today we are focusing on sign language.
- ❖ It is critical to lead a child to the point where language learning can happen. Sensory integration/occupational therapy and dietary interventions are helpful to some.

26

Approaches to Consider
in Facilitating Communication Growth
Autism or Autism-like conditions

- ❖ Concrete System utilizes miniature objects
- ❖ Color coding/Fitzgerald Key – used in gaining structured English language coupled with signing
- ❖ Floortime – building circles of *signed* communication until a level of proficiency is attained
- ❖ ABA or Verbal Behavior reinforcing positive uses of language or behaviors
- ❖ PECS - Picture Exchange Communication System – successful in some people who were considered 'non-verbal'.
- ❖ Apricot - Apricot System – focuses and directs attention on specific happenings by using line drawings

27

Facilitating Communication Growth

- ❖ Observing the child's interests and emotional reactions will provide insights to opportunities to interact, and help you fashion opportunities to learn language.
- ❖ Remember that due to the difficulties, others may avoid communication. You have the golden opportunity to make certain conditions materialize to encourage interactions, and to show the intrinsic value of communicating to the child and to their communication partners.

28

The Key to Unlocking the
Treasure of a Child's Mind

29

is Language



30



PROGRAM SECTION 6

Breakout Session #1
Friday, July 27, 2007, 10:30-11:45
Pacific Ballroom III

Clinical & Genetic
Evaluation of Children
with CHARGE:
The Baylor Study



Presented by
Seema Lalani

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Clinical and Genetic evaluation of children with CHARGE syndrome-Baylor Study
Seema Lalani

Enrollment:

- Most families were enrolled through CHARGE Syndrome Foundation meetings
- Physician referrals
- Children who had Blake's 4 major criteria, 3 major and 3 minor criteria, or 1 or 2 major with several minor criteria, were chosen for the study
- Informed consent was obtained
- Clinical histories were obtained for all probands, as well as the affected family members
- The medical records/photographs were analyzed
- Clinical data available on 134 children

Results:

I) General:

- Mostly sporadic cases
- Seven familial cases
- 66 males and 68 females
- Mutations in *CHD7* gene were found in 86/134 (64%) children

II) Frequency of various problems in children with CHARGE syndrome

Coloboma	83%
Choanal atresia	53%
Deafness	91%
Cleft lip/palate	32%
Facial palsy	54%
Heart defects	81%
Short stature	66%
Swallowing problems	75%
Urogenital problems	65%
Developmental delay	94%
Characteristic CHARGE syndrome ears	99%
Tracheoesophageal fistula/esophageal atresia	17%

III) Important discussion points:

- Most of the affected children have new mutations in the *CHD7* gene.
- Most children with combined coloboma, choanal atresia and inner ear abnormalities have mutations in *CHD7* gene.
- There is no distinct correlation between the severity of the clinical findings and the type of mutations.
- Heart defects, coloboma of the eye, tracheoesophageal fistula and facial asymmetry are also commonly seen in children who have mutations in *CHD7* gene.

IV) Familial cases:

- Rare familial cases with recurrence observed.
- Mutation in *CHD7* gene was identified in 4/7 familial cases. These are:
 - 1) Identical twins: Parents normal
 - 2) Both mother and daughter affected
 - 3) Two affected children- Parents normal
 - 4) Two affected children-Parents normal



PROGRAM SECTION 7

Breakout Session #1
Friday, July 27, 2007, 10:30-11:45
Pacific Ballroom IV

Life, Liberty, and the
Pursuit of Happiness:
Can Emotional
Competence Ever Be an
Educational Goal?



Presented by
David Brown

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

*“Life, liberty, and the
pursuit of happiness”*
Is emotional competence ever
an educational goal?

David Brown
Education Specialist
California Deaf-Blind Services

1

Francis Fukuyama (2002)

“While many would list human reason and human moral choice as the most important unique human characteristics that give our species dignity, I would argue that possession of the full human emotional gamut is at least as important, if not more so”

“Human nature is very plastic ... But it is not infinitely malleable, and the elements that remain constant, particularly our species - typical gamut of emotional responses, constitute a safe harbor that allows us to connect, potentially, with all other human beings.”

2

Without emotions there can be no joy, no values, no motivation. Emotions are very powerful tools for learning. The heart goes out of teaching when we become more concerned with where we’re going than where we and the children are.

Rob Long, Times Educational Supplement 1999

3

EMOTIONAL INTELLIGENCE

- Identifying emotions
- Understanding emotions
- Managing emotions
- Using emotions

Paula Ford-Martin 2002

4

Social and Emotional competence

(Adapted from Jean Brunelli, RN)

SOCIAL COMPETENCE

Behaviors that enable a person to develop, and participate in, positive interactions with others

EMOTIONAL COMPETENCE

The ability to effectively regulate emotions to accomplish your goals

5

Assessing social and emotional development

- There is a sequence to development in both areas
- Development in both is shown by similar activities
- Social development is external, related to functioning with other people
- Emotional development is internal and is demonstrated in self-regulation, focus, and the initiation of both new and familiar activities

6

Key concepts in understanding social and emotional development

- Feeling safe, secure, and understood makes successful learning possible
- The adult-child relationship is the key to the child's resilience
- Physiological, emotional, and behavioral resilience is the key to development
- Nurturing care-giving provides genuine dialogue for the development of attachment & mastery of self-regulation
- Brain research shows lack of "goodness of fit" may lead a child to be prone to distress and to increased regulation problems

7

"Goodness of fit"

- The child's physiological and emotional needs, and the adult's capacity to note and respond to these needs, is demonstrated
- It is critical to brain development, secure attachment, and eventual self-regulation of emotions and behavior
- Occurs easily if the adult has learned to read and respond to the infant's cues, and if the adult's current life circumstances are not overwhelming and provide the necessary social, emotional, and economic support

8

Risk factors

- The child's temperament is intense, cues are sporadic, resistant, or inconsistent
- The above may lead to increased sensitivity, proneness to distress, and self-regulation issues
- Neurological and sensory difficulties may interfere with attachment
- Multiple issues result in altered social and behavioral competencies in both infant and adult
- Social and environmental issues impinge on the infant-adult duo's ability to interact in typical ways

9

[Self-regulation]... "is defined as the capacity to manage one's thoughts, feelings and actions in adaptive and flexible ways across a range of contexts"

Jude Nicholas, CHARGE Accounts, Summer 2007

10

The 9 levels of arousal

(Carolina Record of Individual Behavior)

- Uncontrollable agitation
- Mild agitation
- Fussy awake
- Active awake
- Quiet awake
- Drowsy
- Active sleep
- Quiet sleep
- Deep sleep

11

McInnes & Treffry (1982)

A child who is deaf-blind *may*:

- lack the ability to communicate with his or her environment in a *meaningful* way
- have a *distorted perception* of the world
- be *deprived* of the information necessary to anticipate future events or the results of his or her actions
- be *deprived* of many of the *most basic extrinsic motivations*

12

McInnes & Treffry (1982) cont.

- have medical problems that lead to serious developmental lags
- be mislabeled as developmentally disabled or emotionally disturbed
- be forced to develop unique learning styles to compensate for his or her sensory impairments
- have extreme difficulty in establishing and maintaining interpersonal relationships

13

"Facilitating the Self-determination of Youth and Young Adults with Deaf-Blindness Brian Abery, Deaf-Blind Perspectives, Winter 1998-99

Characteristics of self-determination include the following:

- ✓ An awareness of personal preferences
- ✓ The ability to set goals
- ✓ The ability to use the skills one possesses to achieve goals
- ✓ The ability to evaluate progress toward a goal and learn from experience

14

Lilli Nielsen

"Emotional
development
involves mastery"

15

Deafblind Infants and Children

McInnes & Treffry (1982) p217

The child has:

1. The opportunity to explore the equipment and environmental space in which the activity is to take place
2. An accurate understanding of what is required to complete the task successfully
3. An understanding of how well he/she has performed the task
4. Received communication at the appropriate level to allow him/her to begin to attach labels to actions and objects
5. The opportunity for a constantly enlarging circle of related experience

16

van Dijk & Nelson "Principles of Assessment" (2001)

"The foundation of the assessment is the establishment of a relationship with the child and the creation of a secure base that begins with the child's interests and successes, and it is the responsibility of the assessor to adjust his/her emotions, cognitive level and communications to those of the child." pp 4-5

"Pre-assessment planning begins by talking with the child's parents...to obtain information about the child's history, interests, preferences, motivators...Information is gathered about the child's preferred learning channels...as well as what the child likes and dislikes" pp 5-6

17

van Dijk & Nelson "Principles of Assessment" (2001)

"The child's interests guide the process: therefore, the assessment begins by following the child's interests and movements." p 6

"It is our hope that by carefully following the lead and interests of children...educators will come to know and understand the children and how they learn, communicate, socialize, and solve problems." p 25

18

Natalie Barraga (1976)

Visual functioning is related in part to the condition of the eye. More explicitly, visual functioning is determined by the experiences, motivations, needs and expectations of each individual in relation to whatever visual capacity is available to satisfy curiosity and accomplish activities for personal satisfaction.

19

David Wiley "Where is there joy in this IEP?"
(www.tsbvi.edu/Outreach/seehear/fall04/joy.htm)

- ✓ Find out what is motivating to the student
- ✓ Assess to see if the student is enjoying the day at school
- ✓ Plan activities that incorporate motivating elements for the student
- ✓ Alternate less motivating activities with ones that create joy
- ✓ Teach language using highly motivating topics
- ✓ Teach literacy using highly motivating topics

20

The limits of my
language are the
limits of my world.

Ludwig Wittgenstein

21

Brown 'Follow the Child' (2001)

- Consult those who know the child better
- Identify motivators
- It's okay to match different sensory inputs
- Relax/arouse the child
- Position the child to free up function
- Allow the TIME necessary for the loop of sensory perception>>> interpretation>>> response

22

van Dijk & Nelson "Principles of Assessment" (2001)

- Make the child at ease
- Determine the child's bio-behavioral state
- Determine the child's interest
- Follow the child's interest

23

Jean Ayres' theory

That the environment has a crucial impact on brain development, that the brain changes in response to external stimuli, and that experiences resulting from sensory inputs and the child's responses to them affects brain development.

24

Key principles of Sensory Integration Therapy

1. The Just Right Challenge
2. The Adaptive Response
3. Active Engagement
4. Child-directed

25

SESAME - **S**teps and **E**xcellent **S**trategies for **A**chieving **M**aturity that is **E**mootional

- Be aware of the emotional state of the child - observe, interpret, intervene appropriately
- Consider Social Competence & Emotional Competence inseparable - mastery of skills/ planning & choices/ relationships/ communication
- Consider and deal with the barriers to emotional maturity that are inherent in deaf-blindness
- Explore links between sensory perception and self-awareness of emotional state - consider a Sensory Integration perspective and/or other therapies
- Use communication & language that includes a focus on emotional states right from the start
- Provide appropriate and on-going support for those most involved with supporting the child

26



PROGRAM SECTION 8

Breakout Session #1
Friday, July 27, 2007, 10:30-11:45
Balboa Bay I

Grandparents Session



Facilitator
Nancy Hartshorne

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

08 FS Grandparents Session (*Facilitator: Nancy Hartshorne*)

Grandparents of children with CHARGE syndrome have a unique experience. Grandparents and other adult extended family members are invited to come to this session to share with one another in a safe, facilitator-guided environment. Come and share, or just come and listen. Either way, you are likely to make a connection of support with others living in your shoes.



PROGRAM SECTION 9

Breakout Session #1
Friday, July 27, 2007, 10:30-11:45
Balboa Bay II

Father's Forum:
Fathers in CHARGE



Facilitator
Tim Hartshorne

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

09 FS Father's Forum: Fathers in CHARGE (*Facilitator: Tim Hartshorne*)

Does father know best? In the bewildering world of CHARGE it can be hard for anyone to know best. This forum is an opportunity for fathers to share how they do it; how they figure out how to father a child with CHARGE. The session will be facilitated by Tim Hartshorne. Tim is a psychologist and the father of an 18 year old boy with CHARGE.



PROGRAM SECTION 10

Breakout Session #2
Friday, July 27, 2007, 1:00-2:15
Pacific Ballroom II

Crackerbarrel Session
On Cochlear Implants



Presented by
Ella Taylor,
Kathleen Stremel,
Nancy Hartshorne, and
Gloria Rodriguez-Gil

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

10 MG Crackerbarrel Session on Cochlear Implants (*Ella Taylor, Kathleen Stremel, Nancy Hartshorne, and Gloria Rodriguez*)

This crackerbarrel will serve as a platform in which families can discuss the following:

- 1. Information that was/was not available to them before they made the decision for implantation,*
- 2. The process that they used to make the decision either for or against implantation,*
- 3. The type of intervention/habilitation that their child received or is receiving,*
- 4. Their child's current outcomes, and*
- 5. Their satisfaction with the process (what works, what didn't work, what they would share with other families considering the process).*

We feel that other families, who may be considering implantation, would greatly benefit from this facilitated discussion. Families may also join a LISTSERV for families to increase their networking. The session was designed to achieve the following outcomes:

- 1. Increase in the knowledge of the children with CHARGE who are implanted,*
- 2. Increase in the knowledge of the different types and intensity of habilitation,*
- 3. Increase in the knowledge of family perspectives.*



PROGRAM SECTION 11

Breakout Session #2
Friday, July 27, 2007, 1:00-2:15
Pacific Ballroom III

Transition-to-Adulthood:
Resources and DB-LINK



Presented by
Betsy McGinnity

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007



National Consortium
on Deaf-Blindness

Consortium Members

The Teaching Research Institute
NCDB Central Office
Western Oregon University
345 N Monmouth Ave
Monmouth, OR 97361

Helen Keller National Center
141 Middle Neck Rd
Sands Point, NY 11050

Hilton/Perkins Program
175 N Beacon Street
Watertown, MA 02472

Information

Tel: 800.438.9376
TTY: 800.854.7013
Fax: 503.838.8150
info@nationaldb.org
www.nationaldb.org



Funded through award H220060002 by the
U.S. Department of Education, OSEERS, OSEP.

Betsy L. McGinnity

The National Consortium on Deaf-Blindness (NCDB) is a national technical assistance and dissemination center for children and youth who are deaf-blind. Funded by the U.S. Department of Education's Office of Special Education Programs (OSEP), NCDB builds on the technical assistance activities of NTAC, the information services and dissemination activities of DB-LINK and adds a third focus related to personnel training. NCDB brings together the resources of three agencies with long histories of expertise in the field of deaf-blindness, The Teaching Research Institute (TRI) at Western Oregon University, the Helen Keller National Center (HKNC), and the Hilton/Perkins Program at Perkins School for the Blind. NCDB works collaboratively with families, federal, state and local agencies to provide technical assistance, information and personnel training.

NCDB provides **technical assistance** (TA) and support to assist states in the implementation of IDEA and evidence-based practices. NCDB works to identify the needs of children, their families and their educational teams and implements an array of TA to build the capacity of state and local agencies to meet those needs.

NCDB is home to **DB-LINK**, the largest collection of **information** related to deaf-blindness worldwide. A team of information specialists makes this extensive resource available in response to direct requests, via the NCDB web site, and through topical mailing lists.

NCDB provides leadership in a national effort to promote **personnel training** and professional development. In collaboration with state deaf-blind projects, personnel preparation projects, and other federally funded projects NCDB works to facilitate a variety of personnel training efforts.

For more information about NCDB visit our web site at www.nationaldb.org or contact us at:

National Consortium on Deaf-Blindness
The Teaching Research Institute
Western Oregon University
345 North Monmouth Avenue
Monmouth, OR 97361
(800) 438-9376 Voice
(800) 854-7013 TTY
(503) 838-8150 FAX
info@nationaldb.org

Person Centered Planning

Person Centered Planning is an ongoing problem-solving process used to help people with disabilities plan for their future. In person centered planning, groups of people focus on an individual and that person's vision of what they would like to do in the future.

ARTICLES and OTHER PUBLICATIONS

- **A Brief Guide to Personal Futures Planning Workbook**
 - <http://www.tsbvi.edu/Outreach/deafblind/pcp-workbook.pdf>
 - Texas School for the Blind and Visually Impaired
- **Creating Pathways for Children with Deafblindness - the NTAC/NFADB Parent Training**
 - <http://www.tsbvi.edu/Outreach/seehear/winter04/pathways.htm>
 - Texas School for the Blind and Visually Impaired
- **A Little Book About Person Centered Planning**
 - <http://www.inclusion.com/bklittlebook.html>
 - Inclusion Press
- **The Origins of Person-Centered Planning: A Community of Practice Perspective**
 - http://thechp.syr.edu/PCP_History.pdf
 - Center on Human Policy
- **Person-Centered Planning**
 - <http://challengingbehavior.fmhi.usf.edu/personcentered.htm>
 - The Center for Evidence Based Practice: Young Children With Challenging Behavior
- **Person-Centered Planning: A Tool for Transition**
 - <http://www.ncset.org/publications/viewdesc.asp?id=1431>
 - National Center on Secondary Education and Transition
- **When Planning for Adult Life, How is a "Life-style" Different than a "Program"?**
 - <http://www.tsbvi.edu/Outreach/seehear/winter04/planning.htm>
 - Texas School for the Blind and Visually Impaired

DB-LINK BIBLIOGRAPHIES

- **Person Centered Planning Materials** <http://www.dblink.org/lib/topics/person-centrd-bib.htm>
- **Person Centered Planning Research Materials** <http://www.dblink.org/lib/topics/pcpresearch-bib.htm>

INTERNET RESOURCES

- **Futures Planning and Self Determination**
 - <http://www.partnersinpolicymaking.com/curriculumplanning.html>
 - Partners in Policymaking
- **Person Centered Planning Center of Expertise**
 - <http://www.pacer.org/tatra/personal.htm>
 - Pacer Center
- **Profiles of Courage**
 - <http://www.capacityworks.com/profiles.html>
 - Capacity Works

RESEARCH

- **Building Authentic Visions: How to Support the Focus Person in Person Centered Planning**
 - <http://www.communityinclusion.org/article.php?article%20id=31&staff%20id=42>
 - Institute for Community Inclusion
- **The Most Important Member: Facilitating the Focus Person's Participation in Person Centered Planning**
 - <http://beta.communityinclusion.org/publications/text/rp14text.html>
 - Institute for Community Inclusion
- **Person Centered Planning Research Materials**
 - <http://www.dblink.org/lib/topics/pcpresearch-bib.htm>
 - DB-LINK Bibliography
- **Person Centered Planning Research Overview**
 - <http://www.dblink.org/lib/topics/PCPresearchoverview.htm>
 - National Consortium on Deaf-Blindness
- **Person-Centered Planning: Research, Practice, and Future Directions**
 - <http://www.brookespublishing.com/store/books/holburn-5907>
 - Brookes Publishing

This resource list was excerpted from the Selected Topics Section of the DB-LINK web site. www.nationaldb.org/topics/

July 2007

Sex Education

DB-LINK/NCDB PRODUCTS

- **Introduction to Sexuality Education for Individuals Who Are Deaf-Blind and Significantly Developmentally Delayed**
 - <http://www.dblink.org/pdf/sex-ed.pdf>
 - PDF Format. Requires Adobe Acrobat Reader

ARTICLES and OTHER PUBLICATIONS

- **Opportunities and Barriers for Deafblind People**
 - <http://www.ssc.education.ed.ac.uk/resources/vi&multi/hart97.html>
 - Scottish Sensory Centre
- **Reference Shelf: Sexuality and Puberty**
 - http://www.sesa.org/newsltr/ref_sexuality/slitoc.html
 - SESA
- **Sex education for children with intellectual disabilities**
 - http://www.disability.vic.gov.au/dsonline/dsarticles.nsf/pages/Sex_education_for_children_with_intellectual_disabilities
 - Disability Online
- **Sexuality and Individuals with Developmental Disabilities**
 - <http://www.dddcec.org/positionpapers/SexualityandDD.doc>
 - CEC - Division on Developmental Disability
- **Sexuality and the Deaf-Blind Child**
 - <http://www.sfsu.edu/%7Ecadbs/Summert972.html>
 - California Deaf-Blind Services
- **Sexuality Education for Children with Visual Impairments: A Parents Guide**
 - <http://www.tsbvi.edu/Education/sexuality-education-parent.htm>
 - Texas School for the Blind and Visually Impaired
- **Sexuality in the Context of Personal and Social Development - devising policy and guidelines.**
 - <http://www.ssc.education.ed.ac.uk/resources/vi&multi/jeales98.html>
 - Scottish Sensory Centre
- **Strategies for Minimizing the Risk of Sexual Abuse**
 - <http://www.sfsu.edu/~cadbs/31AbusePrevention.pdf>
 - California Deaf-Blind Services
- **Taking the Fear Out of Sex Education**
 - <http://www.sfsu.edu/%7Ecadbs/Summert97.html>
 - California Deaf-Blind Services
- **Who told you not to do it?": exploring staff attitudes and issues and where conflicts arise for staff**
 - <http://www.ssc.education.ed.ac.uk/resources/vi&multi/phart98.html>
 - Scottish Sensory Centre

DB-LINK BIBLIOGRAPHIES

- **Sex Education Materials** <http://dblink.org/lib/topics/sex-ed.htm>

INTERNET RESOURCES

- **Social Interaction Skills Curricula & Resources**
 - <http://www.tsbvi.edu/bib/social.htm>
 - Texas School for the Blind and Visually Impaired

- **Teaching Tools: Sex Education for Youth with Intellectual Disabilities**

- <http://www.sexualityandu.ca/teachers/tools-10-1.aspx>
- Society of Obstetricians and Gynecologists of Canada

RESEARCH

Sexuality Education for Children with Visual Impairments

- <http://www.tsbvi.edu/Education/sexuality-education.htm>
- Texas School for the Blind and Visually Impaired

This resource list was excerpted from the Selected Topics Section of the DB-LINK web site. www.nationaldb.org/topics/

July 2007



National Consortium
on Deaf-Blindness

Consortium Members

The Teaching Research Institute
NCDB Central Office
Western Oregon University
345 N Monmouth Ave
Monmouth, OR 97361

Helen Keller National Center
141 Middle Neck Rd
Sands Point, NY 11050

Hilton/Perkins Program
175 N Beacon Street
Watertown, MA 02472

Information

Tel: 800.438.9376
TTY: 800.854.7013
Fax: 503.838.8150
info@nationaldb.org
www.nationaldb.org



Funded through award H325060002 by the
U.S. Department of Education, OSERS, OSEP.

Betsy L. McGinnity

Transition Related Materials

Transition

Transition Packet – designed for team members, includes sample fact sheets, articles and resource lists.

Selected Topic Section on web

Full text articles
Checklists and timelines
Links to other resources
Research

Customized Information

Call an information specialist (800) 438-9376
Send an email dblink@tr.wou.edu

Employment

DB-LINK/NCDB PRODUCTS

- Adult Services Referral Checklist for Transition-Age Young Adults Who are Deaf-Blind
<http://www.dblink.org/lib/topics/adultservcheck.htm>
- National Transition Follow-Up Study of Youth Identified as Deafblind: Parent Perspectives
<http://tr.wou.edu/ntac/documents/spotlight/transition.pdf>

ARTICLES and OTHER PUBLICATIONS

- **Assessments of Individuals with Significant Disabilities Under the State Vocational Rehabilitation Services Program**
 - <http://www.ed.gov/policy/speced/guid/rsa/tac-02-01.pdf>
 - United States Department of Education, Office of Special Education and Rehabilitative Services, Rehabilitation Services Administration
- **Developing Employment Opportunities for People who are Deafblind in Victoria**
 - <http://internex.net.au/~dba/papers.htm>
 - Australian National Deafblindness Conference
- **How to Create a Winning Video Resume**
 - <http://www.sfsu.edu/~cadbs/27VideoResume.pdf>
 - CA Deaf-Blind Services
- **MAKING IT Successful Transition Competencies for Youth with Visual Disabilities**
 - <http://www.tsbvi.edu/Outreach/seehear/spring00/makingit.htm>
 - Texas School for the Blind and Visually Impaired
- **Transition From School to Work for People Who Are Deaf-Blind: Some Practical Ideas**
 - <http://www.deafblind.ufl.edu/files/FOPress9.1.pdf>
 - Florida Outreach Press

This resource list was excerpted from the Selected Topics Section of the
DB-LINK web site. www.nationaldb.org/topics/

July 2007



PROGRAM SECTION 11

Breakout Session #2
Friday, July 27, 2007, 1:00-2:15
Pacific Ballroom III

Transition-to-Adulthood:
Taking the Fear Out of
Sex Education



Presented by
Maurice Belote

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

8th International CHARGE Syndrome Conference
Costa Mesa, California
July 27-29, 2007

Taking the Fear Out of Sex Education

by Maurice Belote, Project Coordinator
California Deaf-Blind Services
mbelote@sfsu.edu • www.sfsu.edu/~cadbs

WHAT IS SEX EDUCATION?

Sex education is a comprehensive program that prepares people for satisfying, enriching, and complete lives. It goes far beyond the four subject areas that most adults experienced as children in school: anatomy, physiology, puberty, and reproduction. A comprehensive sex education program has a wider scope, teaching skills such as developing and maintaining friendships, personal and community safety, prevention of abuse and exploitation, physical and emotional intimacy and, most importantly, being in control of your own body. A sex education program provides the information that others need so that they might have the kinds of lives that any of us would wish for ourselves.

WHY IS THIS ISSUE SO IMPORTANT?

Not surprisingly, most of us are uncomfortable talking about sexuality, but ignoring social/sexual issues doesn't make them disappear. No one wins when these issues go ignored. Students are left with misconceptions and sketchy knowledge. Families are left to try to teach these skills at home without the benefit of quality, relevant instructional materials. School and community programs are left with students and clients who may ultimately fail because the system failed them—by not providing them with the information they needed in order to become well-rounded, competent members of society. There is a perception that people with disabilities also have sexualities that are disabled, and this is almost always not the case.

Most people—including children and young adults—learn most of what they know about social relationships and sexuality through incidental learning and through the media (film and television). Individuals with disabilities may not have the advantage of some or all of these modes of learning. For individuals with disabilities who may also have had unsatisfactory past experiences or limited experiences, these social skills—which are often subtle—must be systematically taught. To complicate matters further, issues of sexuality and human relationships are often demonstrated and learned through very subtle means, which may be especially difficult for individuals with sensory impairments such as vision and/or hearing loss. For instance, persons with normal vision and hearing rely heavily on body language, tone of voice, subtle facial expressions, and subtle visual signals when interpreting social situations.

WHY ISN'T SEX EDUCATION TAUGHT IN MOST SCHOOL PROGRAMS?

Most special education programs do not have systematic, planned instruction in the area of sexuality. Programs that do include sex education generally focus instruction for students who are nearing the end of their public school careers (i.e., ages 17 to 21). For students who are fully included in general education programs, the situation is not much better. The standard sex education units that most students receive in public schools (i.e., puberty and menstruation in the upper elementary grades followed by contraception, sexually transmitted diseases, reproduction and childbirth in middle and high school) are frequently not adapted or modified for persons with significant disabilities. These units also do not always include the specific supplemental information required to meet individual needs (e.g., identification and use of the Circle of Friends process, concrete training in purchasing contraception, etc.). Students who are fully included are sometimes simply excluded from sex education instructional programs because school staff members decide that the information is not relevant to students with disabilities.

There is a fundamental paradox in the absence of sex education programs in school programs. When a person with disabilities loses employment, volunteer work, and/or community living arrangements, the reason cited is often related to the person's lack or perceived lack of social (or social/sexual) skills. Even when the cause is not identified as relating to social/sexual skills, needs in this area are sometimes at the root of the problem.

Of course, no one should be forced to teach sex education. It is entirely possible that the subject matter makes some people so uncomfortable that it is not in their best interest (or the interest of the students) to force the issue. Usually there is someone at the school, supported living project, job training project, or community agency who is willing and able to teach sex education.

There seem to be a number of significant barriers to the initiation and year-to-year continuation of sex education programs. While it probably is not necessary or beneficial to dwell too heavily on these issues, identification of these and other barriers will help to facilitate the identification of solutions for overcoming these barriers.

Barrier: We feel embarrassed talking about sexuality. Most of us do not feel comfortable discussing issues related to sexuality, especially with our own children or the students we teach. Many adults were raised with the view that these issues are discussed only in more private settings and under more intimate circumstances.

Barrier: We feel uncomfortable about our own sexuality. It is only natural that we bring a little of ourselves to everything we teach. Issues of sexuality are very personal and, therefore, any discomfort we feel about the topic will likely be evident to the students we serve. Of course, level of comfort is influenced greatly by the way each of us was raised, including cultural factors, religion, family dynamics, and our own positive and negative history of relationships.

Barrier: We do not feel competent to teach the subject. Most of us are not experts on the subject of sexuality, and we may not know where to turn in order to supplement or update our knowledge base. If we depended on teaching what was taught to us, most of us would be significantly hindered in our attempt. For most teachers or agency workers, there is little focus

on the subject of sexuality in higher education unless community colleges and universities offer specific courses that we happen to take as part of the general education requirements.

Barrier: We have a hard time accepting that our own children or the individuals we serve are or will be sexual beings. In essence, we feel uncomfortable about the sexuality of the students (children, youths, and young adults) we serve. This is probably partially due to the fact that we don't have a clear image of what the future holds for these individuals. Not every student who comes to us has had the benefit of a systematic Personal Futures Planning process or MAPS process. Therefore, it is not always clear what skills the students will need in order to succeed. Over-protectiveness on the part of families and/or school staff also affects how we view the future of those we serve.

Barrier: We do not fully understand how other curricular areas relate to sex education. Many curricular areas that are foundations of most school programs are closely connected to many of the goals of a sex education program. These curricular areas include living skills, community instruction, receptive and expressive communication skills, and social skills. It may not be clear to program instructors how easily some of the skills related to social/sexual education can be (or are being) integrated into students' current educational programs.

Barrier: We feel uncomfortable about the required graphic and concrete nature of the instruction. We know that instruction for individuals with significant disabilities (and especially for individuals with vision impairments or who are deaf-blind) must utilize concrete materials and concrete instructional activities. For example, when we want to teach what an apple is, we should not only use a plastic apple or a picture of an apple. We use a real apple that we can smell, touch, taste, chop, cook, etc. This gives accurate information about the real object (i.e., its size, shape, texture, etc.). The use of concrete instructional materials is even more important when the individual has limited success with formal communication systems that might allow for greater use of abstract language.

Barrier: We feel overwhelmed by the breadth of the task. Students may come to a program at the age of 18 without the prerequisite information and skills needed for a sex education program for a typical 18-year old adult. Therefore, there may be a great deal of catch-up instruction required. In this case, the decision of where to begin instruction might seem overwhelming. When this happens, it is sometimes hard to take the first steps to getting started with the program.

WHAT MIGHT BE INCLUDED IN A SEX EDUCATION PROGRAM?

The following represents what might be considered a skeletal outline for a program for all students, including those with significant disabilities.

General development: names and functions of body parts; body changes during puberty; differentiating between babies, children, adolescents, and adults.

Personal safety and community safety: differentiating between familiar people and strangers; inappropriate touching of others and by others; public restroom behavior; assertiveness training; abuse prevention.

Concepts of public and private spaces: identifying personal private places; differentiating between private places in school and community and private places at home; behaviors appropriate to public and private places.

Body care, health, and hygiene: washing and bathing; menstrual care; toileting skills and hand washing; disease prevention; preparation for medical and dental exams.

Affectional expression: appropriate greetings with acquaintances, friends, teachers, etc.; expressions of friendships; choosing and differentiating between friends and acquaintances.

Sexual expression: masturbation; intercourse and birth control; safe alternatives to intercourse; safe vs. unsafe behaviors and sexually transmitted diseases; identification of consensual partners.

Additional course content, depending on specific needs and skills, might include other key areas such as:

- personal values exploration and clarification
- building concepts of self-esteem and positive self-concept
- self-responsibility skills
- problem solving skills
- dating skills
- developing long-term, committed relationships
- pregnancy and family planning
- slang terminology
- alternatives to pregnancy
- child birth
- parenting

WHAT COMPONENTS SHOULD BE CONSIDERED WHEN DESIGNING A PROGRAM?

Relationships built on trust are a critical component to the teacher/student relationship. Learning is most effective when the student determines that his or her teacher is a trustworthy person who will provide accurate information in a safe and positive environment. It is important when choosing instructors for a sex education program that these issues of trust are acknowledged. The subject matter is at times very personal, and it is imperative that the students feel comfortable with the instructor. Conversely, it is also imperative that the instructors feel comfortable with the students. If there is a communication facilitator or interpreter used by the student, it is important that the student feels comfortable discussing these issues through a facilitator. The student must accept the facilitator as a safe person if they are to be willing to discuss personal matters they may have not yet shared with family and/or friends.

Families and educators must work as a team to plan instruction and goals. By working together from the beginning, the team ensures buy-in from all parties involved. This process might begin as one component of the annual family interview or significant other interview. The interview will help the educational staff have a better understanding of family values, as well as the values and standards of the person's local community.

Instruction may require the use of concrete materials and instruction. This is one of the most difficult aspects of sex education for some students with significant disabilities, and is especially true for students with sensory impairments such as deaf-blindness. It is not always possible to "talk around" a subject, or use abstract descriptions to provide instruction. For some students, instruction must be made very real. Obviously, most educational systems in most cultures do not allow the use of real people to teach issues of sexuality. Therefore, it is necessary to make use of models, replicas, etc., and then establish a correspondence between the models and real life. There are companies that sell life-like instructional aids that are either suitable for sex education programs, or that can be adapted without too much trouble. For some instructional purposes, the use of a real object is both possible and preferable. A real condom that can be explored by the group will make a lot more sense than would a picture and verbal description of a condom. In the case of a teaching unit on condoms, it may also be necessary to take additional steps—such as teaching where and how condoms can be purchased or acquired. A trip to a pharmacy, grocery store, or restroom vending machine to locate condoms would be an appropriate way to take the mystery out of an otherwise very mysterious topic.

The program must have administrative support for the purchase and adaptation of instructional materials. Most commercially available family life programs rely heavily on abstract language, visual and auditory learning-- modes that may not be available to all learners. Despite this, the materials and activities in these programs can usually be adapted or modified for individuals with disabilities. By including families and administrators in the planning of sex education programs from the beginning, all parties will understand the importance of the topic, value its inclusion in the school program, and support the need for specialized materials and curricula.

Instruction must be conducted using all appropriate communication modes necessary and using appropriate language levels. The instructors' challenge is to make sure that each student in the program is fully included in the instruction. This means that instruction will need to utilize the modes of communication used by all students in the program—speech, photographs, symbols, raised line drawings, signed communication, voice output systems, pantomime, etc. The instruction will most likely be as individualized and unique as are the individuals themselves.

Instructional objectives must focus on generalization of skills. Instructional activities must be conducted with various materials, in various locations, with various people, and at different times of the day to increase the likelihood that skills will be generalized. For instance, if we teach appropriate restroom behavior and safety, we want to help ensure that the student understands the skills apply in all settings, not just in restrooms familiar to the individual.

A comprehensive evaluation system should be utilized to ensure that skills have been attained and generalized. It is vital for instructors to truly know how thoroughly a skill has been mastered. The students may perform a skill in a particular place with particular people as part of

their daily routine. This same skill may not be exhibited in a less familiar environment or under different or exceptional circumstances.

Instruction should be integrated into all areas of the program. Generally speaking, sex education is conducted during a time set aside for this purpose. This is understandable since the instructors, students, and materials are all assembled in one place. This does not, however, prevent information and skills from being introduced and/or reinforced throughout the rest of the instructional day. This is especially true of times when students are included in instruction and recreational activities with same-age peers who do not have disabilities. By modeling appropriate (and also inappropriate) behavior, same-age peers help build-in intrinsic motivations for students with disabilities. This is especially true if and when a student is at the age when one of her or his primary objectives is to fit in with the rest of the group.

Instruction should recognize the value of the teachable moment. It is much more desirable to teach a skill at a topical moment rather than wait for a contrived setting during a scheduled instructional time. This is especially true for individuals who learn best in natural settings, and this is also when intrinsic motivations are strongest. Teachable moments must be seized, and treasured. Of course, this requires a great deal of flexibility on the part of the instructor—but the benefits cannot be underestimated.

Individual choices must be respected. Inclusion into natural environments with same-age peers who do not have disabilities will provide students more information about all the options available from which to draw on when making choices and decisions. While the choices students make may not be the same choices we as instructors would make, it is our role to validate the choice making process and allow for as much personal choice as possible. As in all other areas of instruction, the instructor may not always be able to honor a particular choice. However, in all cases, the choice needs to be acknowledged and an explanation given as to why the choice cannot be honored at that time. We must also find ways to recognize consumer choices—in whatever way these choices are communicated. This requires that instructors know the formal and/or informal communication systems used by students, or that qualified interpreters/facilitators are utilized to ensure full participation by each individual in the group.

Instruction must recognize the balance between allowing students to make choices and validating choice making while, at the same time, giving boundaries based on current scientific and medical knowledge. This is one of the most difficult aspects to instruction in sex education for individuals who may have developmental disabilities and/or limited formal communication skills. As teachers and family members, we strive to give students factual information, and then guide them in identifying the boundaries of their own behaviors based on individual experiences, attitudes, values, etc. There are, however, sexual practices that put people at risk of contracting life-threatening diseases. The program team must come to agreement on what behaviors will be labeled OK and what behaviors must be labeled as dangerous and "out of the question". These qualifiers must be based on specific individual situations, and by assessing the individual's ability to take precautions, make informed decisions, etc. Of course, the instruction must provide alternative behaviors for the practices that might be ruled "out of the question".

Boys and girls (or young men and women) should not be separated during instruction.

Separation suggests to children and young adults that the "other side" has secrets or mysteries that cannot be divulged. It will make for more natural future interactions if both sexes become accustomed—from the beginning—to discussing issues and raising questions related to sexuality together as a unified group.

Instruction should be provided by a team of at least two persons, preferably by a man and woman working side by side. In terms of professional liability, instruction by two or more people ensures that there is another person present to remember exactly what was taught and discussed. Concern over issues of liability may be an unfortunate reality but one that should not be overlooked. In addition, it also makes good sense to teach in pairs. Team teaching exposes students to different viewpoints and experiences. As much as we strive to keep our own opinions out of the instruction, a topic that is as laden with issues of personal values and beliefs as is sex education is sure to be heavily influenced by what we as instructors bring to the program. When men and women teach sex education together, they will hopefully model positive communication, cooperation between the sexes, and build on and compliment the other's skills.

Instructors can be honest with themselves about their own inadequacies with the subject matter. We are not all experts in the field of sexuality. We all fall somewhere on the continuum, between being very knowledgeable all the way to requiring remedial instruction. One instructor may feel very comfortable with anatomy but not comfortable with talking about feelings and emotions. Another instructor might not be strong in naming the body parts, but may have a gift of leading discussions and/or activities related to values clarification. While it is most desirable that instructors identify personal areas of need and seek out professional growth activities in these areas, it is also fair to acknowledge and take advantage of individual strengths and talents.

Instruction in school, living, or work settings should take into account agency policy statements. These policies, covering issues of sexuality and sexual expression, may already exist or may be at some stage of the development process. It would be unfair for instructors to teach something that would result in an individual being reprimanded for doing what they were taught was acceptable to do. For this reason, it is only fair to students that we assist to inform them of what current policies exist for their home and work settings. In addition, students should know that policies that apply in one location may not apply in others, and that some settings do not fall under policies at all. In the case of the latter, students must make decisions and choices based on their own values, principles, and their understanding of the law.

A sense of humor is a must. Sex education is a difficult subject, and one that makes almost all instructors at least a little anxious. Activities may not always proceed as planned and surprises are inevitable. Maintaining a flexible attitude and the ability to laugh at yourself will help take off some of the rough edges.

[This article has been adapted from an article by the same author that originally appeared in the proceedings of the 1997 National Conference on Deaf-Blindness, *The Individual in a Changing Society*, Washington, D.C.]

Strategies for Minimizing the Risk of Sexual Abuse

by Maurice Belote, CDBS Project Coordinator
mbelote@sfsu.edu • www.sfsu.edu/~cadbs

The incidence of sexual abuse among persons with disabilities is staggeringly high, and yet abuse prevention is rarely addressed in school programs for these individuals. Teaching children who have multiple disabilities including deaf-blindness often requires creativity and the ability to adapt and modify existing materials and programs. When teaching abuse prevention, it may not be adequate to simply follow the same instructional objectives used among children without disabilities (e.g., don't talk to strangers, run away and tell a safe person if someone is trying to hurt you, etc.). For a child who is deaf-blind, intervention will need to encompass many curricular domains, including the areas of communication, self-help, and social skills. The following strategies may be useful in creating an instructional program to address prevention of abuse and exploitation.

Start young. Issues of sexuality begin at an early age, and instruction during these early years creates a foundation onto which everything else can be built. Some of the early skill areas that will assist in abuse prevention instructional activities include curiosity about the bodies of other people (children and adults), names and function of body parts, and public restroom behavior. In addition, this is the time to make children feel comfortable about talking to their parents or caregivers about personal issues. This comfort level—established at an early age—will be very helpful as the child passes through adolescence and young adulthood. Despite what we may think, national research consistently suggests that teenagers *want* to discuss these issues with their parents, and that adult-child communication is effective in decreasing sexual risk behaviors.

Know the people who interact with your child. Sadly, most abusers aren't strangers, but people who know their victims: friends of the family, neighbors, service providers, etc. If a situation doesn't feel right, trust your instincts and intervene. An Internet resource can be found at <http://www.sexoffender.com> that provides a database searchable by state and also a guide to Megan's Law. And while vigilance is important, there is probably no need to be overly suspicious of everyone who interacts with children. The vast majority of friends, neighbors and service providers are caring people who would never put a child's safety and well being at risk.

Make sure skills are generalized. When teaching abuse prevention skills, use the same methods that help ensure that all skills are generalized—teach the skills in multiple locations and settings, with multiple people, and at various times of the day and night. A significant component of skill acquisition is testing to determine if the skill is truly mastered and generalized. Don't assume that a child will perform in a certain way if she or he has demonstrated the skill in a contrived setting with familiar adults. You may need to set up a situation where the child must demonstrate mastery in an unfamiliar setting with unfamiliar people.

Teach terminology, including slang. It may be difficult for an individual to relay information about abuse or mistreatment if the person lacks of way to communicate this clearly. Building vocabulary regarding body parts and action words is an important step to providing the individual with a communication system that will last a lifetime. It may also

be necessary to specifically address the use of slang. For example, an individual who isn't knowledgeable of widely used slang terms for genitalia and sexual acts is more vulnerable because of their lack of sophistication, even if they know the proper "medical" terms for the same things.

Respect privacy; and insist that others do so too. It is important that we provide children with significant disabilities the same respect and dignity we give all people. It may be necessary to teach the concept of modesty, and be certain that this instruction respects individual family values and norms. For children who require help with daily living, issues of privacy and modesty may be complicated by situations where adults and even peers are providing assistance with physical care needs that require intimate physical contact. One way to handle this, from an early age, is to ask the person's permission before helping with intimate or invasive tasks. If requesting permission is established early and consistently, the person who is receiving help is much more likely to feel she or he is in control of their body, and in control of where they are touched and by whom they are touched.

Teach appropriate behaviors. We want to teach our children and students to act in the same way we expect others to when those others interact with our children. For example, we want our children to resist if other people try to touch them in inappropriate places on their bodies. This will be difficult to teach if these same children have been allowed to touch others in those same places. The goal is to establish norms, so that behaviors outside of these norms are clearly viewed as such.

Put it in the IEP. Don't assume that goals and objectives discussed in the IEP meeting will be implemented if they are not part of the written plan. It isn't necessary to include *everything* in an IEP, but too often there is a reluctance to include items in IEPs that are out of the ordinary domains such as functional academics, gross/fine motor, communication, etc. If a particular skill is very important to you, do not accept an explanation that instruction in this skill doesn't need to be written into the IEP because it will be addressed all the time throughout the child's program. The IEP is the family's assurance that a skill will be addressed, and also provides a forum for discussing mastery towards the goal at subsequent IEP and team meetings. For service providers, IEPs provide concrete plans, and help maintain consistency between programs and staff members during times of transition or instability.

Sexuality Education

Web Resources for Serving for Children and Youth who are Deaf-Blind and/or have Significant Disabilities

www.siecus.org

The Sexuality Information and Education Council of the U.S. (SIECUS)

SIECUS is a national, nonprofit organization which affirms that sexuality is a natural and healthy part of living. Incorporated in 1964, SIECUS develops, collects, and disseminates information, promotes comprehensive education about sexuality, and advocates the right of individuals to make responsible sexual choices.

www.sexualhealth.com

The Sexual Health Network

The Sexual Health Network is dedicated to providing easy access to sexuality information, education, mutual support, counseling, therapy, healthcare, products and other resources for people with disabilities, illness, or natural changes throughout the lifecycle and those who love them or care for them.

www.tr.wou.edu/DBLINK/lib/products.htm

Introduction to Sexuality Education for Individuals Who Are Deaf-Blind and Significantly Developmentally Delayed, by Kate Moss and Robbie Blaha. Monmouth, Oregon: DB-LINK, 2001. A book for parents and professionals that offers information and instructional guidance for delivering sex education to deaf-blind students who also have cognitive disabilities. Issues of self-expression related to gender identity, modesty, and appropriate touch are discussed. Specific instruction is included for menstruation, masturbation, hygiene, health, and sexual abuse. This document also available on CDROM.

<http://www.sfsu.edu/~cadbs/News.html>

Summer 1997 issue of California Deaf-Blind Newsletter *reSources* on the topic of sexuality education.

www.sfsu.edu/~cadbs/English.html

California Deaf-Blind Services Fact Sheet #31: *Strategies for Minimizing the Risk of Sexual Abuse*

www.diverse-city.com

Diverse City Press Inc.

Publisher and distributor of instructional books, videos and DVDs—including “No! How!!!”—specific to teaching individuals with developmental disabilities.

www.stanfield.com

James Stanfield Publishing Company

Comprehensive multi-media instructional materials including the “Life Horizons” series (set includes slides and discussion guides) and the “First Impressions” series on personal hygiene.

www.teach-a-bodies.com

Teach-A-Bodies

Anatomically-correct teaching figures.

www.jimjacksonanatomymodels.com

Reproductive models including models of genitalia, reproductive systems, and pregnant uterus.



PROGRAM SECTION 12

Breakout Session #2
Friday, July 27, 2007, 1:00-2:15
Pacific Ballroom IV

CHARGE 101 for New Families

CHARGE Syndrome Diagnosis
and Features

Presented by

Meg Hefner



Anesthesia Complications

Presented by

Kim Blake

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

CHARGE Syndrome diagnosis and features

Meg Hefner, M.S.
Genetic Counselor
Clinical Associate Professor of Pediatrics
St. Louis University School of Medicine

Where does the name come from?

**1981 Pagon, et al.

- C = coloboma of the eye
- H = heart anomalies
- A = atresia of the choanae
- R = retardation of growth or development
- G = genitourinary anomalies
- E = ear anomalies and/or deafness

**No longer used for diagnosis

Epidemiology of CHARGE Syndrome

- 1 in 10,000 births
- Most often a new dominant mutation
 - Advanced paternal age
 - Increased but low recurrence risk
 - Major gene identified 2004 (CHD7)
- Mild end of spectrum is unknown
 - Parents or other family members with “suspicious” findings

Recurrence

- Empiric risk of 1% for parents with one child with CHARGE
- 50% for children of individuals with CHARGE



1998/2003 CHARGE
Diagnostic Criteria
Major and Minor Features

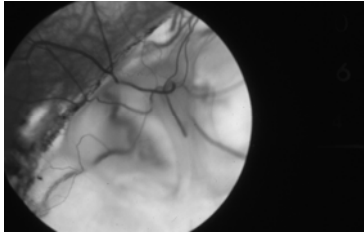
Definite CHARGE Syndrome:

- 3 or 4 Major Features
- 2 Major Features + 3 Minor Features
- Positive gene test (CHD7)

CHARGE Syndrome
Major Features (the 4 C's)

- Coloboma of the globe
- Cranial nerve anomalies
- Choanal atresia/stenosis
- Characteristic CHARGE ears

Coloboma – cleft of the eye



Retinal Coloboma
Increased risk of retinal detachment with retinal coloboma



Iris Coloboma

Retinal coloboma causes upper visual field defects



Macular coloboma – central vision loss

- The child will look above the point of interest and not appear to be making eye contact



CHARGE Syndrome Major Features (4 C's)

- Coloboma of the globe
- Cranial nerve anomalies
- Choanal atresia/stenosis
- Characteristic CHARGE ears

Cranial nerves 9 & 10: Swallowing problems



- Many require G-tube feeding
- May last for years
- Some adults still avoid certain textures

Swallowing complication: Aspiration

- Most common reason for hospitalization in first 2 yr. of life
- GE reflux is common
 - Surgery (Nissen) may help
- Gastrostomy may be needed to reduce aspiration

Cranial Nerve # 7 Facial palsy



Unilateral –lopsided face



Bilateral – no facial expression

Cranial Nerve #1 Sense of smell

Decreased or absent sense of smell

- Difficult to evaluate, but EXTREMELY common (>90%)
- Changes “taste”
- Implications for feeding
- Social implications
 - smelly feet
 - body odor
 - Passing gas

CHARGE Syndrome

Major Features (4 C's)

- Coloboma of the globe
- Cranial nerve anomalies
- Choanal atresia/stenosis
- Characteristic CHARGE ears

Choanal atresia/stenosis

- Two sides or one side
- Bony or membranous
- Blockage (atresia) or narrowing (stenosis)
(One-sided stenosis may be difficult to diagnose)
- Often requires several surgeries to remain open



Baby with stents

Choanal stenosis

- Narrowing of nasal passages
- Goopy nose all the time
- Lots of ear infections, fluctuating hearing



CHARGE Syndrome Major Features (4 C's)

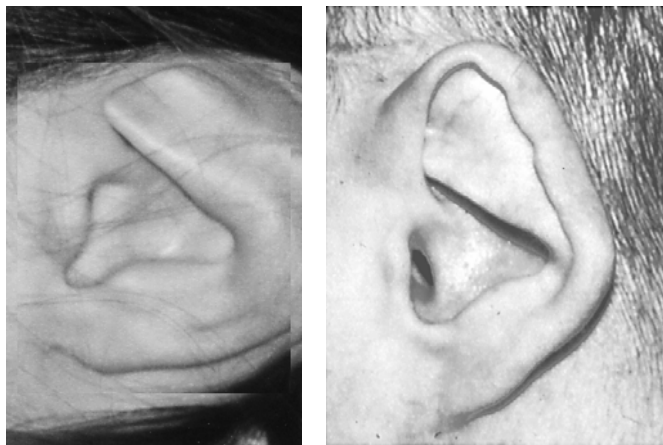
- Coloboma of the globe
- Cranial nerve anomalies
- Choanal atresia/stenosis
- Characteristic CHARGE ears

Characteristic CHARGE Ear: OUTER EAR

- Asymmetry between the two ears
- Floppy (deficient cartilage)
- Small/absent earlobe
- Triangular concha
- Clipped off helix (outer fold)

Characteristic CHARGE Ear

- Short, wide, triangular concha,
absent lobe



Characteristic CHARGE Ear

Snipped off helix with
small or absent
lobe



Discontinuity between the
antihelix and antitragus

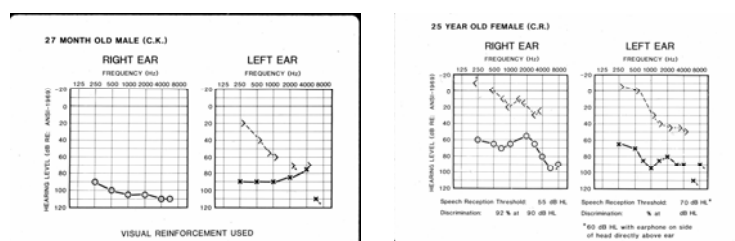


Characteristic CHARGE Ear: MIDDLE AND INNER EAR

- Middle ear bones – ossicular malformations (stapes, incus)
 - Conductive hearing loss
- Inner ear
 - Mondini defect: 1-1/2 turns to the cochlea
 - Small or absent semicircular canals
 - Balance problems

Hearing in CHARGE

- Mixed hearing loss
- Fluctuating with infections
- May respond to cochlear implant
- May be progressive



Inner Ear Balance Problems

- Small or absent semicircular canals
- Characteristic gross motor development
 - Stay close to the ground
 - Creep or combat crawl
 - 5-point crawl
 - “Bottom shuffle”
- Age of walking
 - Walk at 24-32 mo. if no other problems
 - Walk at 3-4 yr. if hypotonia and visual impairment

CHARGE Syndrome Minor Features

- Some are very common to CHARGE but difficult to evaluate consistently
- Some are very common to CHARGE but also very common in other syndromes
- Some are less common in CHARGE

CHARGE Syndrome Minor Features Unique to CHARGE

- Upper body (?truncal) hypotonia
- Characteristic CHARGE face
- Hockey stick palmar crease

Truncal hypotonia

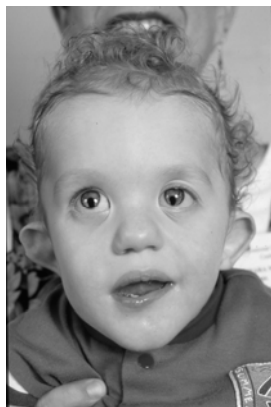
- Slumping posture
- Crawl on back
- Bottom shuffle
- Delayed walking
- Complicated by poor balance



Characteristic CHARGE Face

- Square face
- Broad forehead
- Round eyes
- Nose
 - Prominent bridge
 - Square root
 - Prominent columella
- Small chin, gets bigger with age

Characteristic CHARGE Face



To see more faces: "About CHARGE" link at www.chargesyndrome.org

CHARGE: 7 to 17 years



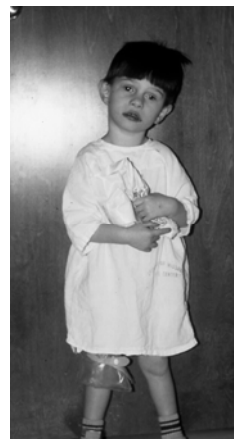
Minor feature: Hockey-stick palmar crease



CHARGE Syndrome

Minor Feature: Heart defects

- Tetralogy of Fallot
- Aortic arch anomalies
- Complex heart defects
- Can be any heart defect
- Same spectrum of heart defects as in VCFS (22q deletion syndrome)



Heart surgery at four years old

CHARGE Syndrome

Other Minor Feature: Clefts

- Cleft lip
- Cleft lip and palate
- Cleft palate
- Submucous cleft palate



Repaired cleft lip

CHARGE syndrome minor feature: Esophagus/Trachea

- Esophageal atresia (EA)
 - esophagus not connected to stomach
- Tracheo-esophageal fistula (TEF)
 - connection between trachea and esophagus
- H-shaped TEF
 - can be hard to diagnose, but important
- Trachoemalacia
 - weak, collapsing trachea

CHARGE syndrome minor feature: Renal (kidney) anomalies

- Hydronephrosis
- Reflux
- Horseshoe kidney
- Small or absent kidney

- 40% have renal anomalies

CHARGE Syndrome Minor Feature: short stature

Due to:

- Medical problems
 - Heart
 - Feeding
- Growth hormone deficiency
- Short stature with no known cause



Treated with growth hormone

CHARGE Syndrome

Minor Feature: genital hypoplasia

- Males
 - Micropenis
 - Cryptorchidism (undescended testes)
 - Lack of normal puberty
- Females
 - Small labia
 - Absent uterus
 - Lack of normal puberty



17 year old with no hormone treatment (and facial palsy)

- EVERY feature can vary from absent to severe in different children
- NO ONE FEATURE is seen in every person with CHARGE
- Definite CHARGE is 3 or 4 Majors or 2 Majors and 3 Minors
- Probable/Possible CHARGE is more difficult
- The new gene test may help, but \$\$

Beyond diagnosis: Other features (any organ system!)

- Brain anomalies seen on MRI or CT
- Seizures
- Apnea
- Laryngomalacia
- Floppy cartilage
- Nipple anomalies
- Thymic or parathyroid abnormalities

More Other features

- Webbed neck
 - Sloping shoulders
 - Absent trapezius muscle
- Abdominal wall defects
 - Umbilical hernia
 - Omphalocele
- Limb/skeletal anomalies
 - Polydactyly common in Japan
 - Scoliosis

CHARGE management issues beyond Major and Minor features

- Constipation - Autonomic nervous system?
- Potty training – nerve abnormalities?
- Sleep disturbances
 - abnormal circadian cycle
- Cyclic vomiting/abdominal migraines
- Sensory integration
- Behavior

Behavior in CHARGE syndrome

- Autistic-like behaviors
 - May be deafblind behavior
 - May be autism
 - May be CHARGE
- Stubborn, perseverative
- OCD
 - A real feature of CHARGE

Intelligence in CHARGE

- Long-term prognosis may be excellent
- Most are “input impaired” due to combined vision & hearing loss
- A few are “output impaired” due to bilateral facial palsy
- Delayed motor milestones due to vestibular dysfunction, upper body hypotonia and impaired vision.

How well do they do?

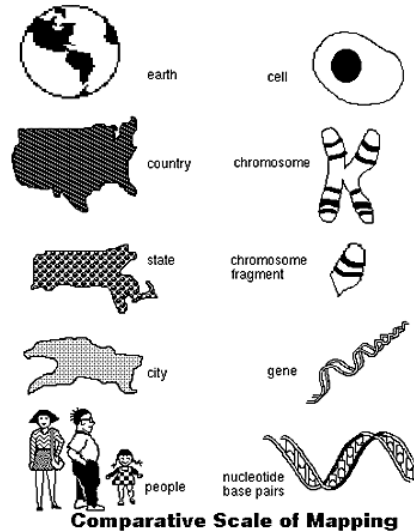
The typical 2 year old with CHARGE:

- has spent 6 months in the hospital
- has had at least 6 surgeries
- is followed by 10 medical specialists
- is fed by G-tube
- is not walking or talking
- has some unusual behaviors

Looks pretty “retarded,” but probably isn’t

A major gene for CHARGE

Finding a change (mutation) in a single gene is like locating a single person from space



Major CHARGE gene: CHD7 on chromosome 8q12

- Very large gene
- Many, many different mutations have been identified
 - Makes testing very expensive
- CHD7 is a regulatory gene (turns other genes on and off): Thus affects every organ system

Now what - How is this gene helpful?

- Confirm diagnosis in questionable cases
 - Help define the mild end of the spectrum
 - Not finding a mutation does NOT rule out CHARGE
- Test other people in the family
 - Can “normal” people be carriers?
- Prenatal diagnosis
 - First must identify the mutation in the family
 - Can identify the gene, not the severity of the features

ANESTHESIA COMPLICATIONS IN CHARGE SYNDROME

Dr. Kim Blake, MD, MRCP, FRCPC



**8TH International CHARGE Syndrome Conference
July 27 – 29, 2007
Costa Mesa, California**

There are Always Risks of Complications with Anaesthesia

- “...you sign a consent”
- Are you informed?

- Are Individuals with CHARGE Syndrome More at Risk?

If yes, what are the risks and who should know?



APSEA 2006

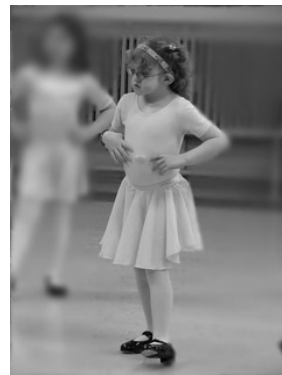
Growing up With CHARGE Syndrome



Age 0-2 years: 7 surgeries



Age 2-4 years: 3 surgeries



Age 4-6 years: 6 surgeries

Kennedy

Kennedy's ICU Admissions 4 in Total

- 5-19 days – open heart surgery and supraventricular tachycardia.
- 6 months – G-tube/fundoplication
- extubation attempted (x 3)
- 18 months – aspiration pneumonia
- 6 yrs – post op heart surgery – 1 day ? pneumonia

Frederick's Story



Freddy - Early Days

- Difficulty with intubation
- ToF repair, vascular ring repair, PDA ligation
- ↑ secretions
- Difficulty with extubation

Botox was Used for Increased Oral Secretions

Drooling, excessive secretions (sialorrhea)

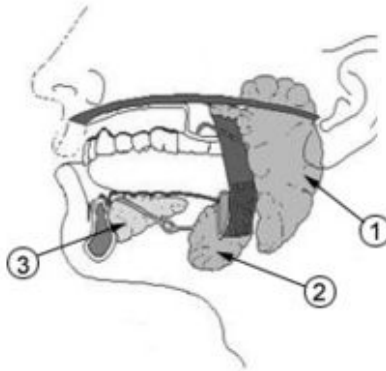
- Infrequent swallowing
- Ineffective swallowing

Can be related to neurological conditions

?cranial nerve anomalies

Site of Botox Injections

1. Parotid glands
2. Submandibular glands
3. Sublingual glands



Freddy at 9 Months

- Viral pneumonia
- Aspiration pneumonia
- Gastroesophageal reflux

- Re-injection of botox

STUDY: Detailed chart review: 4 females, 5 males, average age 11.8 yrs

- Surgeries (ears, diagnostic, digestive/feeding, nose, throat, dental, heart, eyes, other)
- Anesthesia type/number
- Complications – major (reintubation NICU admission, minor (post-op cough, wheeze, crackles)

Results (9 individuals)

- 218 surgeries (average of 24 per child!)
- 147 anesthetics (16 per child)
- Average age of first operation: 8.8 months (3 days to 4 years)
- Average surgeries per individual: 21.9 (+/- 12.2)

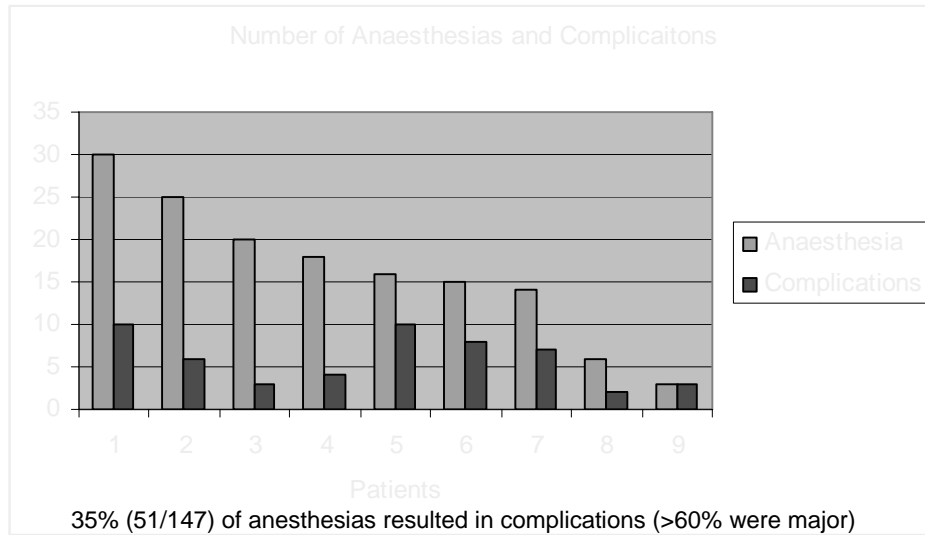
Results

Type of Procedure	Number of Procedures	% of Total
Ears	47	22
Diagnostic	44	20
Digestive/Feeding	31	14
Nose/Throat	30	14
Dental	26	12
Heart	20	9
Eyes	6	3
Other	14	6
Mean length of anesthesia 124 minutes (+/- 31.6 minutes)		

Single vs Multiple Procedures

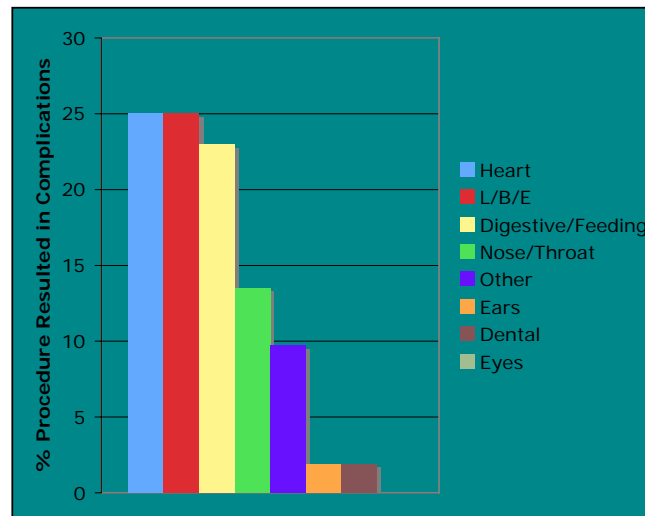
Single	Multiple
39%	27%
37/94	14/51
P>0.05	

Results



Results

Anesthesia related complications occurred most often with heart, diagnostic scopes and gastrointestinal tract procedures



MacKenzie's Story



- 27 surgical procedures
- 18 anaesthesias
- 4 complications
- Multiple ICU admissions

Discussion

- 35% of anesthetics resulted in complications
- Heart, diagnostic, and gastrointestinal tract procedures result in the most complications
- A complication resulted at least once in every type of surgery except for eyes

Discussion

- High risk of complications in individuals with Nissen fundoplication or gastrotomy/jejunostomy tube
- Low risk with cleft palate
- What about individuals with CHD7 mutations who have mild clinical criteria?
 - Will they be at risk in the future?
 - Have they actually been challenged with surgeries?

Conclusion

Individuals with CHARGE are at high risk of anesthesia complications, especially post operatively. Combining procedures during one anesthesia does not increase the risk of anesthesia-related complications. The anesthetist needs to be aware that, even with simple procedures, the individual with CHARGE syndrome is at high risk of complications.



PROGRAM SECTION 13

Breakout Session #3
Friday, July 27, 2007, 2:30-3:45
Pacific Ballroom I

STRATEGIC CONFLICT
MANAGEMENT SKILLS IN
SPECIAL EDUCATION
SETTINGS



Presented by
John Reiman

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

*CADRE is the OSEP-funded National Center on Dispute Resolution in Special Education.

The materials presented herein are adapted
from presentations made nationwide by
CADRE.

1

Today's Objectives

- ▣ Participants will gain awareness of 'listening to understand' as an essential relationship, communication, coalition-building and advocacy tool.
- ▣ Participants will gain awareness of the difference between 'positions' and 'interests' – and how this difference powerfully impacts coalition-building, communication and advocacy.
- ▣ Participants will, through discussion, relate workshop content to their own experiences.

2



Effective Communication?

(from reSources: CA Deaf-Blind Services, Winter '06 – written by Alexis's mother)

Amy (deaf-blind student) and Alexis (intervener) have been a successful team for 12 years now. They have progressed through 4 schools, 14 teachers, numerous psychologists and therapists. Unfortunately, throughout all these years, Alexis has not always had the benefit of supportive mentors or administrators. Often her role as intervener has been misunderstood, and she has had to fend off attempts to encroach upon her commitment to Amy. This has been awkward at times. Sometimes teachers haven't known how to direct Alexis' efforts or how to use her expertise to develop appropriate instructional strategies for Amy.

4

Effective Communication (cont'd)?

Some underestimate the value of an intervener's role and resent the input of a non-credentialed employee, no matter how well reasoned or accurate. Some have abandoned their responsibility to Amy and left her to Alexis' care and instruction. This lack of support has sometimes left Alexis fatigued and disillusioned. Through the years, despite her own professional woes, Alexis has never betrayed Amy's trust or stopped advocating for her needs.

**POINT: HOW MUCH COMMUNICATION HAS THERE BEEN?
MIGHT THERE BE A ROLE FOR > COMMUNICATIVE,
RELATIONAL, AND PROBLEM SOLVING SKILLS?**

5

Assumptions...

- ▣ Different cultures have differing perspectives on conflict and how it is approached
- ▣ Conflict is a healthy reflection of a diverse and changing society
- ▣ Most parent/school relationships are or can be positive and mutually respectful
- ▣ Skills can be acquired for productive relationships (personnel prep and parents nationwide)

6

Assumptions...

- ❑ Early opportunities exist to collaboratively address differing viewpoints on how to best serve a child
- ❑ Parents should know laws/regulations for advocacy – **and** adversarial processes should be reserved for situations where other options have been exhausted
- ❑ The cost of adversarial processes, *when necessary*, can be high in personal, relationship and financial costs
- ❑ It's not hard to end up "in the drink" when people don't listen to or understand each other.

7



CADRE/IDEA Partnerships



CADRE/IDEA Partnerships



CADRE/IDEA Partnerships



CADRE/IDEA Partnerships



CADRE/IDEA Partnerships



CADRE/IDEA Partnerships

Listening to Understand

Instruction:

Identify a situation/issue that you have/had strong thoughts and feelings about, and are comfortable sharing here today (pick manageable issue.)

18

CADRE/IDEA Partnerships

High Risk Responses (listening?)

1. Ordering	7. Judging
2. Threatening	8. Praising
3. Moralizing	9. Name Calling
4. Advice	10. Diagnosing
5. Logical Argument	11. Reassuring
6. Questions	12. Diverting

19

CADRE/IDEA Partnerships

Certain Responses ...

- ❑ Derail the conversation
- ❑ Take the focus off the other
- ❑ Block the other from finding a solution
- ❑ Lower the other's self-esteem
- ❑ Distance you from the other
- ❑ Diminish the other's motivation

20

CADRE/IDEA Partnerships

Listening to Understand

Following the thoughts and feelings of others to understand what they are saying from their perspective, frame of reference, or point of view.

Dignity and Respect

21

CADRE/IDEA Partnerships

Listening ⇨ Problem Solving

- ❑ Sometimes speakers just need to be heard
- ❑ Sometimes listening clears up confusion
- ❑ Sometimes listening identifies a need for problem solving

22

CADRE/IDEA Partnerships

Listening to Understand (Exercise Set-Up)

- ❑ Pair up. Choose "A" and "B".
- ❑ "A" – Identify a situation/issue that you have/had strong thoughts and feelings about, and are comfortable sharing here today (pick manageable issue.)
- ❑ "B" – Seek to understand what is powerful in the situation for "A" as a human being, and communicate your understanding to "A."

23

CADRE/IDEA Partnerships

Listening to Understand (Do)

- ❑ Maintain curious inquiring mind about "A"
- ❑ Put personal opinions, beliefs, and judgments about "A's" situation aside – not relevant
- ❑ Find the heart of "A's" experience by listening for what's 'behind' the situation. Listen to understand "A," not the situation.
- ❑ Use silence . . . wait
- ❑ Let "A" know you "get" what is personally powerful for him/her from the situation
- ❑ Name the EMOTION you believe that "A" is experiencing

24

Listening to Understand (Avoid)

- ❑ Interrupting
- ❑ Debating or trying to fix it for "A"
- ❑ Sliding opinion in under guise of listening
- ❑ 4 C's: **Contesting, Convincing, Convicting or Converting**
- ❑ Rehearsing response – it will come
- ❑ Too many questions = not listening
- ❑ Confusing yourself with "A" (projection makes perception)

25

Exercise (beyond paraphrasing and reflection)

1. "A" -- Share situation succinctly (2 minutes)
2. "B" -- Feedback your **"reach for understanding"**: Use expressions like, "Sounds like...." Read between the lines (1 minute)
3. "A" - Correct or validate "B's" attempt to "know what you mean." Share more of situation. Educate "B" so "B" can "get it" fully.
4. "B" -- Feedback more **'reach for understanding'**
5. Repeat Steps 3 & 4 until "A" reports **feeling heard and understood**.

26

Positions & Interests

27

Positions & Interests

Position

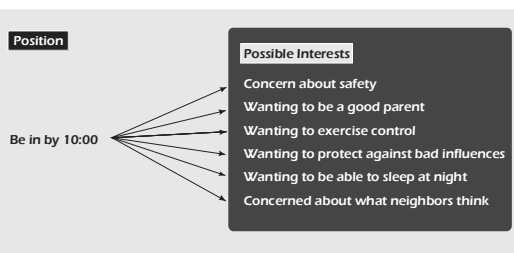
Specific solution proposed to resolve problem - the "WHAT"

Interest

Underlying real need/desire that gives position its life (e.g., beliefs, expectations, values, fears, priorities, hopes, concerns) – the "WHY"

28

Positions & Interests



29



Sample Positions:

- ❑ *"The target level for appropriate interactions in my classroom must remain at 50%."*
- ❑ *"We want an American Sign Language interpreter in that English Lit class."*
- ❑ *"I demand an apology now!"*
- ❑ *"Rob has a right to a full time instructional assistant next term"*

31

Finding the Interests

Question, question, question...

- ❑ *"Why is that solution so important for you?"*
- ❑ *"Why are you suggesting...?"*
- ❑ *"What would you accomplish in getting what you want?"*
- ❑ *"What if that did/didn't happen?"*
- ❑ *"How will you be affected by...?"*
- ❑ *"Imagine that you got _____; what would be taken care of?"*

32

Finding the Interests

- ❑ What need is the person taking this position attempting to satisfy?
- ❑ What is motivating the person?
- ❑ What is the person trying to accomplish?
- ❑ What is the person afraid will happen if a demand is not fulfilled?

33

Demonstration

Identify participants' positions and interests.

34

Summary: Interest-Based Negotiation

- ❑ Aims not to change the other person, but to change negotiation behavior
- ❑ Shifts from "your position versus mine" to "you and I versus the problem"
- ❑ Involves a mutual exploration of interests to yield more creative options
- ❑ Uses objective criteria

Adapted from Highnam, K. (2001). Interest-based negotiation. CSSEA 2001 Fall Conference and AGM. Surry B.C, Canada. CSSEA; Fisher and Ury, *Getting to Yes*.

35

Working Effectively with Schools and Other Service Providers

- ❑ Take someone along
- ❑ Listen
- ❑ Use the power of silence
- ❑ Positions/interests
- ❑ Evaluate compromises
- ❑ The power of "I don't know what you are talking about"
- ❑ "If this were your child..."
- ❑ Prepare questions
- ❑ Get involved
- ❑ Acknowledge good work

36

Setting up a Peer Support Group

- ❑ Informal/formal – electronic/face-to-face
- ❑ Listen to understand -- hard experiences – confidentiality
- ❑ Identification of shared challenges
- ❑ Celebrations of triumphs
- ❑ Network beyond local
- ❑ Write (*DBP*)

37

To listen a soul into disclosure and discovery is the greatest service one human can do for another.

Quaker saying

38

Contact Info

John W. Reiman, Ph.D.
Associate Director, National Consortium on
Deaf-Blindness
Mediation Specialist, CADRE*

reimanj@wou.edu
503-838-8776
503-838-8821 tty
504-838-8150 (fax)

39



PROGRAM SECTION 14

Breakout Session #3
Friday, July 27, 2007, 2:30-3:45
Pacific Ballroom II

Familial CHARGE Syndrome:
Is there a recurrence risk?
Presented by
Conny van Ravenswaaij & Jorieke Bergman



Puberty & Smell in CHARGE
Presented by
Jorieke Bergman & Conny van Ravenswaaij

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Familial CHARGE syndrome; is there a recurrence risk?

Since 2004 we know that CHARGE syndrome is caused by a change in the CHD7 gene. Every person has **two** CHD7 genes, one inherited from father and the other one inherited from mother. CHD7 is a regulatory gene. It regulates the expression of developmental genes very early during the development of the unborn child (embryo). If there is insufficient CHD7 the risk for developmental defects in specific organs like the heart, eye, ear, kidney, etcetera, is elevated (figure 1). A change in **one** of the two CHD7-genes is enough to result in CHARGE syndrome. However, CHARGE syndrome is highly variable and it is not possible to predict the clinical consequences for the child from the specific change in CHD7.

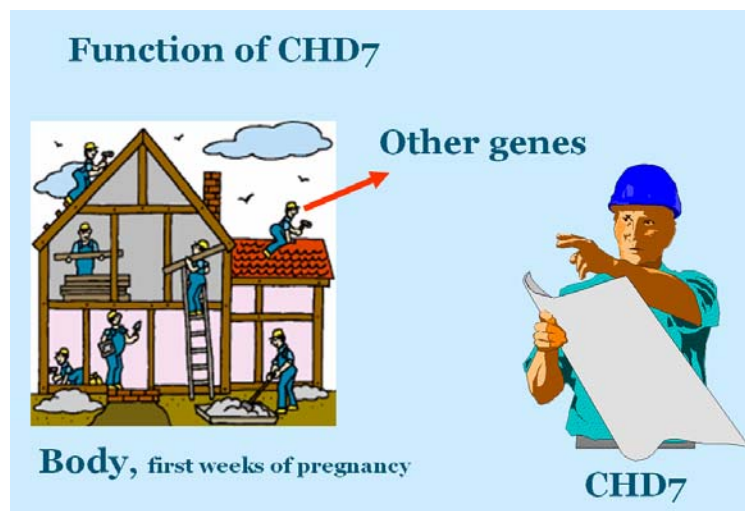


Fig. 1 CHD7 regulates the function of developmental genes during early pregnancy.

Two children with CHARGE syndrome within one family incidentally occur. CHARGE syndrome has a birth prevalence of approximately 1 in 10,000 newborns. About 3% of all persons with CHARGE syndrome have a sib or parent who also has CHARGE syndrome. We have collected as much as possible information of such families because this reveals important information on the variability of CHARGE syndrome. Within families all affected family members have the same change in CHD7, while their clinical problems may differ a lot. Moreover, we discovered that within families often an unexpectedly mild form of CHARGE syndrome can be found. In such families mildly affected persons were identified through a child with typical CHARGE syndrome.

In my presentation I will focus on why CHARGE syndrome sometimes occurs in more than one family member and what the risk of recurrence is.

Monozygotic (identical) twins

CHARGE syndrome is almost always caused by a new change in CHD7. This means that the change occurred in the single egg or sperm cell that gave rise to the child. Thus the change is present in the fertilized egg. This fertilized egg will start dividing, billions of times, and in all cells that arise from the fertilized egg the CHD7 change will be present. Thus the CHD7 change will be present in all cells of the embryo, and subsequently in all cells of the child (figure 2).

If the early embryo divides into two embryos (monozygotic or identical twins), both children will have the same CHD7 change and thus they will both have CHARGE syndrome.

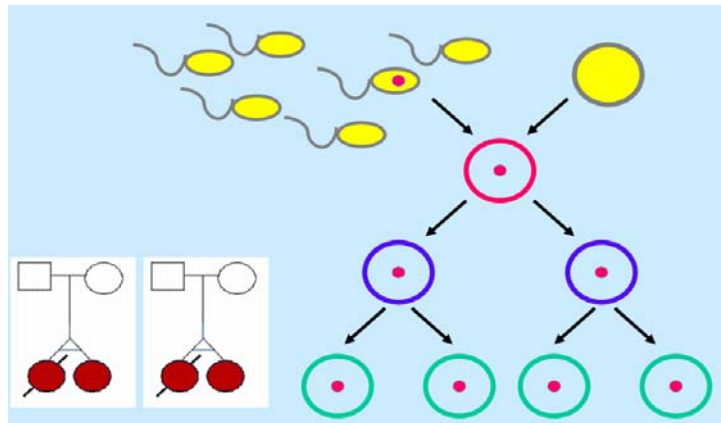


Fig.2 An egg is fertilized by a sperm cell containing a change in the CHD7 gene. All cells that arise from this fertilized egg will carry the CHD7 change. If two children develop from one fertilized egg they will share all genetic information and thus, in this situation, both will have CHARGE syndrome. The pedigrees show two of our families with monozygotic (identical) twins.

A parent with CHARGE syndrome

We collected four families with a mildly affected parent and one or two children with mild to moderate CHARGE syndrome (Figure 3). In all families the diagnosis was made initially in a child with clear symptoms of CHARGE syndrome.

The mildest consequences of a CHD7 change appeared to be abnormal shape of the ears and balance disturbances (abnormality of the semicircular canals). DNA studies of the CHD7 gene could confirm the diagnosis in the parent and, of course, the affected child has the same CHD7 change as the parent.

In these families the recurrence risk is 50% for each pregnancy. Parents always give half of their genetic information to their children. The parent with CHARGE syndrome thus passes on either the normal CHD7 gene or the changed CHD7 gene. This explains the 50% recurrence risk.

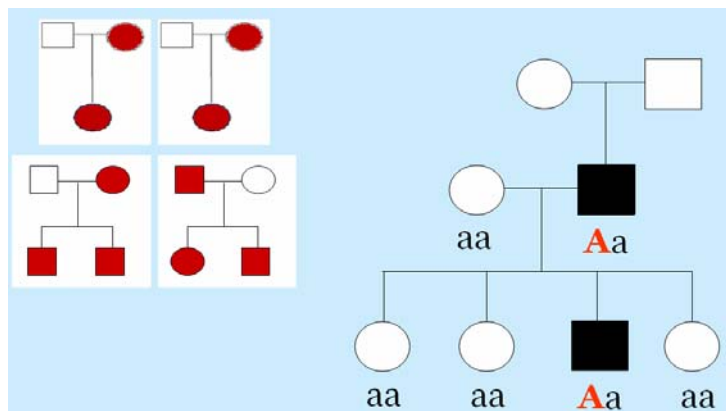


Fig. 3 Four families with parent-to-child transmission of CHARGE syndrome. The parent with (mild) CHARGE syndrome passes on the normal CHD7 gene (a) or the changed CHD7 gene (A). Thus the recurrence risk is 50% for each pregnancy.

Two children with CHARGE syndrome and healthy parents

In five families two sibs of different ages had CHARGE syndrome. Within each family the same CHD7 change was found in both affected children. Thus recurrence had occurred while the parents did not show any symptom of CHARGE syndrome. How is that possible?

In two of these families we showed that one of the parents carried the CHD7 genes in part of his/her body cells. This is called a mosaic. A mosaic situation can occur if in the fertilized egg no CHD7 change is present, but this change occurs after a few cell divisions (figure 4). Only the cells that arise from the cell with the altered CHD7 gene will carry this change. If these cells are also present in the ovaries or testes, egg or sperm cells with the CHD7 change can be formed. If egg or sperm cells with a CHD7 change are involved in fertilisation, a child with (non-mosaic) CHARGE syndrome will be born (figure 2).

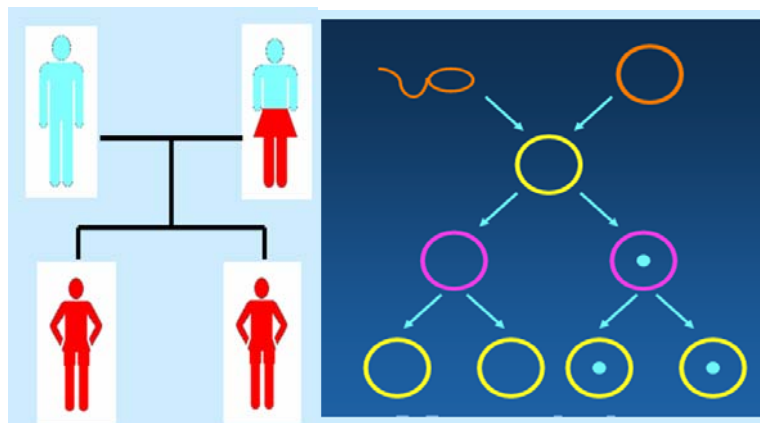


Fig. 4 In this example mother has a CHD7 change in part of her body cells (mosaic). The scheme on the right side explains that this can occur when the CHD7 change arises during a cell division *after* fertilization. Mother will not have CHARGE syndrome herself, but she has an increased risk for children with CHARGE syndrome (she can pass on the CHD7 change via an egg cell, and subsequently the child will have the CHD7 change in all cells).

In the other three families we did not reveal a mosaic in one of the parents. Nevertheless their affected children carried the same CHD7 change. The most likely explanation is that a low-grade mosaic is present in the ovaries or testes, but not in other body cells (or in such a low percentage that this cannot be detected by DNA studies). This is called a germline mosaicism.

What is the recurrence risk in CHARGE syndrome?

Familial CHARGE syndrome is extremely rare. This already demonstrates that in general the recurrence risk will be low. If parents have a child with CHARGE syndrome and want to be informed about future pregnancies we recommend the following:

- Investigate parents for mild symptoms of CHARGE syndrome (hearing, balance, smell, shape of the ears).
- If a CHD7 change has been found in the child, offer DNA analysis to the parents as well.

If the CHD7 change can be found in the parent, either in mosaic or all cells, than the recurrence risk is elevated (maximal 50%). If the parents want to, prenatal diagnosis can be performed in future pregnancies. However one should be aware that the severity of CHARGE syndrome can not be predicted by DNA-analysis.

If the CHD7 change can not be found in one of the parents (the most frequent situation), than a small risk for germline mosaicism remains. Therefore the recurrence risk is not zero, but 1 to 2 %. Also parents without CHD7 change may opt for prenatal diagnosis if they want to.

As mentioned before, the severity of CHARGE syndrome cannot be predicted by DNA-analysis. Fetal ultrasound examination can give extra information, for example on the presence of a heart defect. But not everything can be seen by ultrasound, for instance deafness, developmental delay and behavioural problems will remain undetected. The choice for prenatal diagnosis will always remain a personal one, and the task and challenge of the clinical geneticist/genetic counsellor is to inform the parents in such a way that they can make the choice that fits them best.

Dr Conny van Ravenswaaij, MD PhD
Clinical geneticist
University Medical Center Groningen
c.m.a.van.ravenswaaij@medgen.umcg.nl

Smell and puberty in children with CHARGE syndrome

Introduction

In CHARGE syndrome many congenital abnormalities can occur. Well known features are coloboma, heart defects, atresia of choanae (blockade of the passage from nose to mouth), retardation of growth and development, deafness and ear anomalies. A less well known feature of CHARGE syndrome is the inability (or diminished ability) to smell. Also puberty development can be delayed or absent in people with CHARGE syndrome. These two features seem independent from each other, but they are linked as I will show below.

Smell and puberty

CHARGE syndrome is not the only disease where smell and puberty disorders occur together. Kallmann syndrome is another example. In this disease anosmia and delayed puberty are the main features. Other associated problems (which are only occasionally present) are tooth abnormalities, an absent kidney, cleft lip or palate and mirror movements of the arms (this means that if you move one arm, the other arm involuntarily 'mirrors' this movement).

How do we smell?

Olfaction happens in the nose. Odors are detected by the olfactory neurons which bundle to form the olfactory nerve (first cranial nerve). Eventually the odor information reaches the brain (the olfactory cortex), where we become aware of the smell. Odor information is stored in the long term memory (hippocampus and limbic system) and is connected to our emotional memory and behavior.

Timing of onset of puberty

There is wide variation in the age at which puberty begins. For example, in girls puberty starts earlier than in boys, and Asians are later than African Americans in this respect. The onset of puberty is affected by both genetic and environmental factors. Puberty is a gradual process which starts in the brain. The initial signal that puberty is about to begin comes from the hypothalamus. The hypothalamus releases pulses of GnRH (Gonadotropin-Releasing Hormone) which stimulates the pituitary gland (also located in the brain). The pituitary gland on its turn releases the hormones LH (Luteinizing Hormone) and FSH (Follicle Stimulating Hormone). These hormones, also called gonadotropins, are secreted in the blood and circulate through the body. The ovaries of a girl react on the rising amount of LH and FSH with the production of estrogen, the female sex steroid. Estrogen initiates puberty in girls, characterized by a growth spurt, pubic and axillary hair growth, breast development and menstruation (and fertility). In boys the testes respond to the elevated levels of LH and FSH and they start producing testosterone, the male sex steroid. Under the influence of testosterone puberty is initiated. Boys will undergo height growth, hair growth (pubic, axillary and facial hair), voice change and increase in testicular and penile size (and fertility).

Link between smell and puberty

The link is located in the brain. During early embryogenesis, the olfactory system and the hypothalamus develop together. When the development of the olfactory system is disturbed, you often see a developmental defect of the hypothalamus as well. That is why smell and puberty defects frequently occur together.

Smell test

It is possible to test the sense of smell with a test. At our CHARGE outpatient clinic we use the UPSIT. UPSIT is an abbreviation for University of Pennsylvania Smell Identification Test. The test is composed of 4 booklets, each with 10 questions. Every page contains microencapsulated odorants which are released by scratching. The questions are multiple choice, with four possible answers. For example, this odor smells like: gasoline, pizza, peanuts or lilac. In total 40 odors are tested and the number of correctly identified odors can

be compared with children of the same age and sex. Then we are able to tell if a child is anosmic (unable to smell) or hyposmic (has a decreased sense of smell). All CHARGE patients tested by us so far (nine) have anosmia.

Puberty development

At the CHARGE outpatient clinic a pediatric endocrinologist (a pediatrician specialized in hormones) is present to score the pubertal stages. On physical examination several pubertal features are scored, among which are pubic and axillary hair growth, breast development and testicular and penile size. In addition the hormonal changes can be measured in the blood.

CHARGE: smell and puberty

Clearly smell and puberty are linked, but what does this have to do with CHARGE syndrome? *CHD7* (the gene involved in CHARGE syndrome) seems to have an effect on the development of the brain. In particular, we think that a mutation in *CHD7* might lead to a disturbed development of the olfactory system and the hypothalamus in the brain. In addition *CHD7* has several other effects, for example on the developing heart, the ears and the eyes.

Ongoing research

We wonder if anosmia or hyposmia might be predictive for puberty problems. So we asked ourselves if we can predict with a smell test whether a child will or will not enter puberty spontaneously. In order to answer this question, we are performing smell tests in children with CHARGE syndrome at our outpatient clinic and we follow their puberty development.

The importance of smell

Smell is probably the most underrated sensory organ. Only when we are temporarily devoid of our sense of smell (for example during a cold), then we truly appreciate it. Smell is important because it has an alarm function. Think about leaking gas or rotten food for example. In addition our smell amplifies our sense of taste. The human tongue can only distinguish about five types of taste, while the nose can detect hundreds of odors even in small quantities. Without smell we would not know the difference between an apple and a potato. Also, smell is important in social interactions. Smell makes us aware of our own perspiration and it is also linked to our memories.

Unfortunately a treatment for anosmia is currently not available. However, if you are aware that you have no sense of smell you can take preventive measures (for example buy a smoke detector, profusely use deodorants, etc.).

Effects of delayed puberty

Delayed puberty is very inconvenient for a child. Peers will notice that the child has not entered puberty and might start teasing. Another aspect is that delayed puberty can give rise to osteoporosis. The sex steroids estrogen and testosterone are important for mineralization of our bones. When these hormones are not sufficiently produced, bones can get under mineralized, which makes them prone to fractures.

Delayed puberty can be treated with hormones.

Conclusion

A defective sense of smell and delayed puberty are frequent features of CHARGE syndrome. These features occur because mutations in the *CHD7* gene presumably give rise to disturbed development of the olfactory system and the hypothalamus. We expect that a smell test can predict whether a child with CHARGE syndrome will undergo spontaneous puberty or not. Treatment of anosmia is not possible, but preventive measures are important. Delayed puberty can be treated with hormones in order to prevent osteoporosis.

Ms. Jorieke Bergman, MD PhD-student
Clinical geneticist in training

University Medical Center Groningen
j.e.h.bergman@medgen.umcg.nl

Definitions and abbreviations:

Puberty: process of physical changes by which a child's body becomes an adult body capable of reproduction.

Olfaction: sense of smell, the ability to detect chemicals (odors) dissolved in air.

Anosmia: inability to smell

Hyposmia: decreased ability to smell

GnRH: gonadotropin-releasing hormone

LH: luteinizing hormone

FSH: follicle stimulating hormone



PROGRAM SECTION 15

Breakout Session #4
Friday, July 27, 2007, 2:30-3:45
Pacific Ballroom III

Transition-to-Adulthood:
Glimmers of Promise



Presented by
Joe McNulty
and Kathy McNulty

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Transition to Adulthood: Glimmers of Promise



Joe McNulty, HKNC
Kathy McNulty, NCDB
CHARGE Conference 2007
Costa Mesas, CA



Transition & Adult Services Today

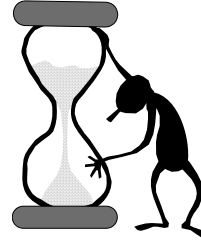


To better understand the present and plan for the future, it is often helpful to look back on what came before...

HKNC & NCDB 2007

A Quick Look Back

- Pre 1920's:
 - little federal presence;
 - institutional settings
- 1920-30's
 - impact of WW I
- 1940's
 - impact of WW II; Rehabilitation Act;
- 1950's
 - parent movement; Arc established
- 1960's
 - Kennedy Era & increased awareness; Medicaid enacted 1965; Elementary & Secondary Education Act of 1965; HKNC Act



HKNC & NCDB 2007

A Quick Look Back

- 1970's – Deinstitutionalization & mainstreaming
- Rehabilitation Act of 1973
 - mandated services for persons with severe disabilities;
 - Status 26
 - Competitive
 - Sheltered Workshops
 - Home maker
 - Independent Living Centers established;
- 1975 Education of All Handicapped Children Act (EHA)
 - Free Appropriate Public Education (FAPE)
 - Least Restrictive Environment (LRE)
 - IEP, due process, integration



HKNC & NCDB 2007

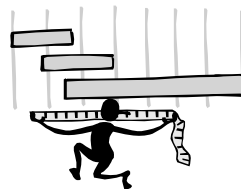
A Quick Look Back



- 1980's – Community, Integrated Settings
 - first Medicaid waivers
 - supported employment demonstration projects funded
 - Developmental Disabilities and Bill of Rights Act 1984
 - vocational focus; functional curriculum; school to work, community-based instruction
 - Person-centered planning introduced
- 1990's - Inclusion
 - consumer choice in career opportunities;
 - IDEA & transition services; inclusion; access to general curriculum; assistive technology devices
- 2000's – Accountability/Evidence-based practices
 - NCLB 2001; IDEA 2004;
 - WIA & One Stop; competitive employment only
 - Department of Labor – added focus on significant disabilities

HKNC & NCDB 2007

Progress made...



- Awareness is dramatically different
- Services have improved and are more appropriate
- As a field we are raising expectations!!

HKNC & NCDB 2007

But!!!

- Shortage of qualified personnel is an on-going challenge.
- Adequate funding to ensure appropriate services for individuals with disabilities is still needed.
- Little attention is given to the development and funding of programs that support the family unit.
- The need continues to increase the awareness and knowledge of parents, service providers and policy makers on effective practices that will lead to successful transition outcomes.



HKNC & NCDB 2007

Glimmers of Promise

NCLB & IDEA

- Impact on instruction & transition services
 - Accountability & assessments
 - State standards and accessing general education curriculum
 - Teaching academics & functional life skills
 - Summary of Performance Exit Report



HKNC & NCDB 2007

Glimmers of Promise

Research on Teaching Self-Determination & Relevance to Transition Services

- By definition components of self-determination include:
 - Choice making
 - Consider options
 - Problem solving
 - Identifying interests, preferences & strengths
 - Initiate
 - Negotiate
 - Set goals
 - Regulate behavior
 - Compromise
 - Persistence
 - Pride
 - Self-confidence



HKNC & NCDB 2007



Glimmers of Promise

NEW MODELS

- Partnership with Community Colleges (CC)
 - Massachusetts Inclusive Concurrent Enrollment Partnership: high school students enroll at CC; state program jointly supported by special ed dollars & CC; HS & CC apply jointly for this money;
- Customized Employment Initiative
 - Dept of Labor – important, new initiative to employ persons with significant disabilities
 - Office of Disability Employment policy (ODEP) favorable results from an independent evaluation
 - Importance of the Discovery Phase
- Independence/New Direction Waiver
 - Self-directed; individualized budget; person-centered plan

HKNC & NCDB 2007

Glimmers of Promise

- **Workforce Investment Act**
 - Rehab Act
 - Transition Amendment, new language
- **Technical Assistance Circular on Eligibility**
- **Secondary Transition State Capacity Building Initiative (OSER, SEA, & State VR)**
- **Technology**
 - Cochlear Implants
 - Screen Braille Communicator
 - Video Relay System



HKNC & NCDB 2007

Suggestions

- Start early
- Stay informed
- Talk to other parents – find examples
- Create a vision
- Learn Person-Centered Planning techniques
- Encourage a team approach
- Use their forms, use their language e.g. exit forms
- Practice and acquire advocacy skills
- Find a vital person!
 - Good at what they do
 - Busy
 - Know a lot of people
 - Nice
- Don't awfulize!! (Loretta Roche)

HKNC & NCDB 2007

Summary of Performance Exit Report

PEORIA UNIFIED SCHOOL DISTRICT NO. 11
PO BOX 39, PEORIA, ARIZONA 85380
TELEPHONE (623) 486-6000

Student information

Date completed: _____

Student: _____ Date of birth: _____ Year of graduation/exit: _____

Evaluation date: _____ Primary disability: _____ Secondary disabilities: _____ / _____

IEP date: _____ Case manager: _____ Phone: _____

Student's desired post secondary goals:

Employment: I would like to:

Education: I will:

Training: I will:

Independent living: I see myself living:

Summary of performance: (complete all that are relevant to the student)

ACADEMIC	Present level of performance (grade level, standard scores/date, strengths, needs)	<u>Essential</u> accommodations, assistive technology, or modifications utilized in high school, and why needed
Reading		
Math		
Written Language		
Other: (note taking, time management, study skills, learning style, etc.)		

FUNCTIONAL AREAS	Present level of performance (strengths and needs)	Essential accommodations, modifications and/or assistive technology utilized in high school, and why needed
Social skills and behavior		
Independent living skills		
Environmental access/mobility		
Communication		
Self-determination/ self-advocacy skills		
Vocational/ transition/ employment		
Other important considerations to assist in making decisions about disability determination and needed accommodations		

Recommendations to assist the student in meeting postsecondary goals: (modifications, accommodations, adult services)
--

WHERE TO FIND INFORMATION ON TRANSITION TO ADULT SERVICES CHARGE CONFERENCE 2007

DEAF-BLIND RESOURCES

Helen Keller National Center
www.hknc.org

National Consortium on Deaf-Blindness (NCDB)
www.nationaldb.org

DB-LINK (NCDB)
Information & Dissemination on Deaf-Blindness
www.nationaldb.org
1-800-438-9376

TRANSITION RESOURCES

National Center on Secondary Education and Transition
<http://www.ncset.org>

National Secondary Transition Technical Assistance Center
www.nsttac.org

Partnerships with Community Colleges
http://finance1.doe.mass.edu/grants/grants07/rfp/doc/235_b.pdf
Initiative being conducted in Massachusetts

A Guide for Students Who Are Deaf-Blind Considering College
www.hknc.org

Project Teams
Frequently Asked Questions about Transition:
<http://www.projectteams.org/transition.php>

DEVELOPMENTAL DISABILITIES/MEDICAID RESOURCES

National Association of Directors of Developmental Disabilities
<http://www.nasddds.org>

To read about a New Directions Waiver click on
<http://www.tash.org/mdnewdirections/ndwaiver.htm>

You can view the latest summary report of all regular approved home and community-based services waivers at:
<http://www.cms.gov/medicaid/1915c/regular.pdf>

The latest summary report of all model approved home and community-based services waivers is at:
<http://www.cms.gov/medicaid/1915c/model.pdf>

Institute for Community Inclusion
National Center on Workforce and Disability/Youth
<http://www.communityinclusion.org>

A Guide to Medicaid and Medicaid Waivers
<http://www.arcind.org/guide> to Medicaid and Medicaid w.htm
Although developed by Indiana, much of the information is for general use.
Medicaid Waivers
<http://www.dpw.state.pa.us?Disable/MentalRetardationServices/ServiceProg/003670156.htm>
Developed by Pennsylvania, information has general use.

SELF-DETERMINATION RESOURCES

The Center for Self-Determination

www.self-determination.com

Click on Links for a listing of additional self-determination resources

PERSON-CENTERED PLANNING

Introduction to Person-Centered Planning

<http://www.ilr.cornell.edu/edi/pcp/courses.html>

PACER Center – Person Centered Planning

<http://www.pacer.org/tatra/personal.htm>

Listing of additional on-line resources provided.

CUSTOMIZED EMPLOYMENT RESOURCES

Fact Sheet on Customized Employment

<http://worksupport.com/resources/viewContent.cfm/502>

<http://www.dol.gov/odep/pubs/custom/index.htm>

ASSESSMENT/ALTERNATE ASSESSMENT MATERIALS

National Center for Learning Disabilities

<http://www.ncld.org/content/view/290/322>

Brief on Determining Appropriate Assessment Accommodations for Student with Disabilities

ADVOCACY/LEGISLATION RESOURCES

National Center on Low-Incidence Disabilities

<http://nclid.unco.edu/newclid/Resources.php>

Pop-Up IEPs, advocacy tips

Wrightslaw

<http://www.wrightslaw.com/idea>

Law & Regulations

The Arc

<http://capwiz.com/thearc/home/>

Action Center~ Elected Officials~ Issues& Legislation

Medicaid updates

Council for Exceptional Children

<http://www.cec.sped.org>

Policy & Advocacy; Legislation Action Center

The Individuals with Disabilities Education Improvement Act of 2004

Office of Special Education Programs (OSEP) Web site,

<http://www.ed.gov/about/offices/list/osers/osep>

IDEA

<http://www.nichcy.org/idea.htm>

Training materials on the law & IDEA regulations

Family Voices

<http://www.familyvoices.org>

Legislative Action Center

FAPE

www.fape.org

IDEA/Laws

PACER

www.pacer.org

Legislative & transition info



PROGRAM SECTION 16

Breakout Session #3
Friday, July 27, 2007, 2:30-3:45
Pacific Ballroom IV

CHARGE 102 for New Families
Feeding in CHARGE Syndrome

Presented by
Kim Blake



Physical Influences on
Development in CHARGE

Presented by
Sandra Davenport

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Feeding in CHARGE Syndrome

Dr. Kim Blake, MD, MSc, FRCPC



**8TH International CHARGE Syndrome Conference
July 27 – 29, 2007
Costa Mesa, California**

Feeding Difficulty in Children with CHARGE Syndrome

- High prevalence of feeding difficulty early in life
 - High incidence of tube feeding (90%)
- Persistent difficulties into adolescence
 - Difficulty managing food textures

Feeding Issues

- Impaired coordination of jaw, tongue and palate movements involved in chewing and swallowing
 - weak sucking
 - nasal regurgitation, gagging, choking
 - aspiration

Feeding Issues

- Reduced sensory awareness in the mouth, palate and throat
- Gastroesophageal reflux
- Reduced appetite
- Abnormal sense of smell
- Learned aversion or refusal

Major Factors Contributing to Feeding Problems

- ❖ Primary is cranial nerve anomalies:
 - CN V, VII, IX, and X
- Cardiovascular difficulties early in life
- Structural anomalies:
 - Choanal atresia/stenosis
 - Tracheoesophageal fistula, esophageal atresia
 - Laryngomalacia, laryngeal clefts, and short aryepiglottic folds
 - Cleft lip/palate
- Behavioural issues

- Feedings disorders associated with CHARGE syndrome may initially have an organic or underlying medical or neurological basis, but may be maintained over time by remaining sensory and oral motor skill deficits as well as the acquisition of maladaptive behavioral patterns.

Recommendations

- Referral to a pediatric gastroenterology specialist in the neonatal period, as soon as the diagnosis of CHARGE is made
- Follow-up should involve a multidisciplinary feeding team
- Parents should be made aware of the potential for long-term feeding issues.

Dobbelsteyn C, Marche DM, **Blake KD**, Rashid M. 2005. Dysphagia. 2005, Vol. 20: 89-100.

MacKenzie & Kennedy: Best Feeding Friends



Recent Research

Feeding difficulties in children with CHARGE syndrome: prevalence, risk factors and prognosis (Dobbelsteyn, Peacocke, Blake et al Dysphagia)

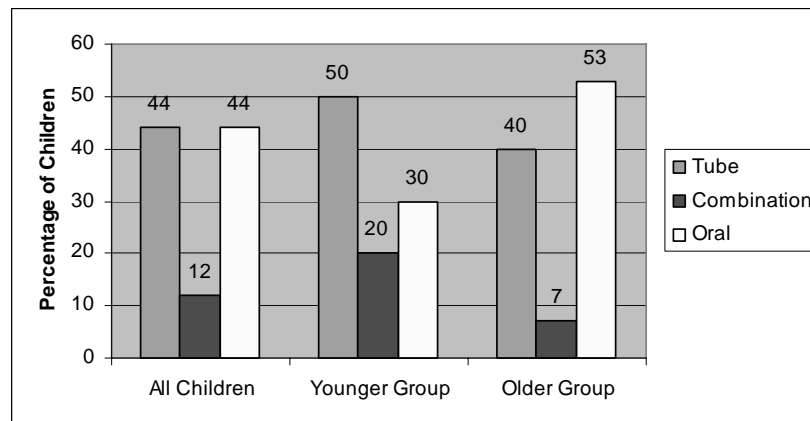
- Investigated the prevalence of feeding difficulty and tube feeding in 39 children with CHARGE syndrome
 - Over 90% of children were tube fed at some time
 - Most children were either exclusively tube fed or totally orally fed
 - CN dysfunction was implicated in feeding difficulties

Follow-up Study

(Dagley & Dobbelsteyn, 2006)

- Investigated changes in the development of oral feeding in Peacocke's population of 25 children (aged 2y4m to 16y5m) over a two-year period
 - Attempted to describe the period of transition from tube to oral feeding

Prevalence of children receiving various types of feeding



Number of children experiencing feeding issues at the two points in time

	2004	2006
Weak chewing	19	12
Weak sucking	19	9
Reduced sense of touch	11	8
Swallowing difficulty	15	14
Aspiration	5	10
Gastroesophageal reflux	11	12
Facial palsy and weakness	14	12

Number of children experiencing feeding issues at the two points in time

	2004	2006
Choanal atresia/stenosis	7	9
Reduced sense of smell	1	5
Hypersensitivity to touch	11	10
Poor control of tongue	2	10
Reduced sense of taste	0	8
Heightened sense of taste	0	2

Further Results

- Children receiving a combination of oral and tube feeding
 - All children showed an increase in the variety of foods consumed orally compared to two years earlier
 - All children still showed some difficulty with thickened liquids

- Children who were exclusively orally fed
 - The majority of parents (78%) reported an increase in their child's willingness to accept a variety of different textures
 - 67% of the children experienced difficulty drinking thin liquids
 - All children were still experiencing difficulty drinking thin and thickened liquids

Had feeding abilities improved over the past two years, and how had they changed?

- Higher measures on the scores obtained for the entire sample indicates improvement in feeding abilities
- 63% of the parents felt that their child's feeding had improved
- Greater number of children were receiving exclusive oral feeding in 2006 compared to 2004
- Older children were more likely to be exclusively orally fed suggesting that feeding difficulties reduce with age

What factors may be attributed to these changes?

- Improved chewing 20%
- Improved sucking 27%
- Better control of tongue 53%
- Improved swallowing 53%
- Decreased aspiration
27%
- Decreased reflux 27%
- Reduced sensitivity 47%

What factors may be attributed to these changes?

- Incr tolerance of textures 40%
- Incr tolerance of utensils 53%
- Incr motivation/
interest in eating 67%
- Increased hunger 47%
- Increased age 67%
- Improved health 53%
- Improved general devel 73%

What was the transition from tube to oral feeding like?

- The average age of introduction of oral feeding for those who were once exclusively tube fed was 3 years of age

Introduction of oral feeding was associated with

- assistance of a feeding team 100%
- improved health 60%
- decreased aspiration 60%
- improved swallowing 60%
- and increased interest in eating 40%

For children who were once exclusively tube fed

- The duration of time from introduction of oral to removal of the feeding tube ranged from 1 year to 7 years
 - All children experienced some setbacks in oral feeding including food refusal, gagging on food, and spitting out food and liquid

Greatest Increase in Score

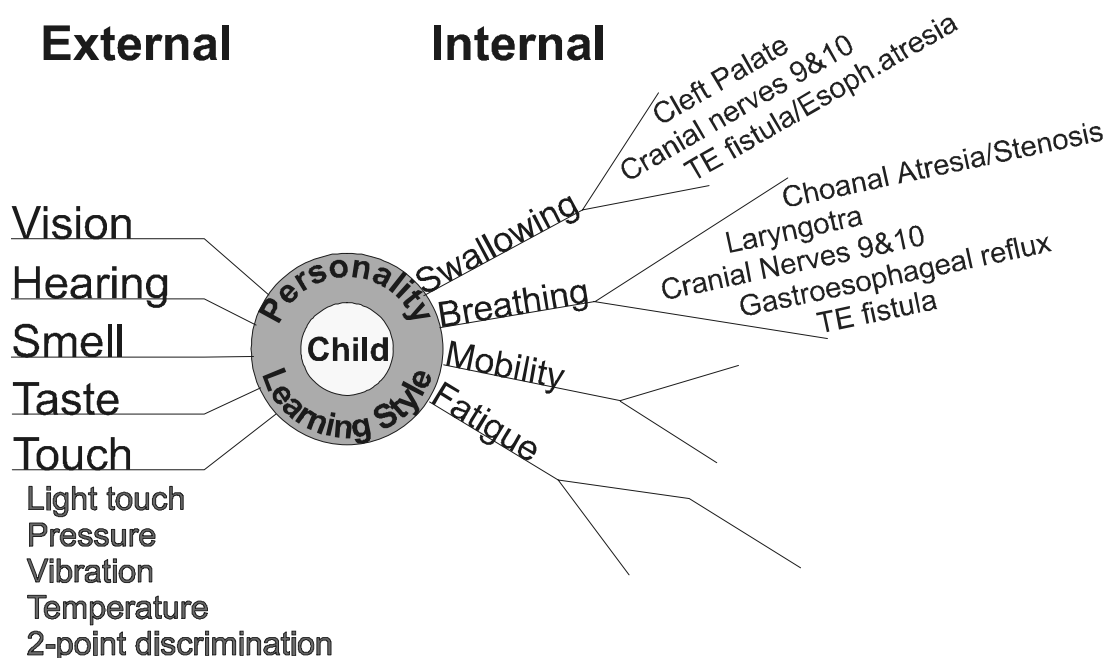
- Male, age 3y9m
- Increase from PASSFP score of 8 in 2004 to score of 31 in 2006
- Improved from “not safe to feed orally” to “some oral feeding with modifications required”
- Persistent feeding difficulties included:
 - weak chewing and sucking, difficulty with taste, facial weakness, poor control of tongue movement and swallowing difficulties, and aspiration difficulties
- Gastroesophageal reflux has been a persistent problem for this child

Greatest Decrease in Score

- Female, age 10y5m
- Decrease from a score of 36 in 2004 to a score of 6 in 2006
- Declined from “able to do some oral feeding with special modifications” to “not safe to feed orally”
- Parent report from 2004 indicated that she was completely fed by mouth around the time she entered school
- In 2006, oral intake of food was completely eliminated
- Reflux was found to be a persistent and prevalent problem for this child in 2006

CHARGE 102: PHYSICAL INFLUENCES ON DEVELOPMENT IN CHARGE

Sandra L.H. Davenport, M.D.
SensoryGenetic/Neuro-development,
5801 Southwood Drive, Bloomington MN 55437-1739
952-831-5522 slhdaven@tc.umn.edu



By Sandra L.H. Davenport, M.D.

CHARGE is such a complex disorder that many of the physical components have a profound effect on the child's development and understanding of the world. The above diagram breaks these influences down into those that are External and those that are Internal. In one way all are internal since the five senses are all part of the body; however, these senses require outside stimuli in order to perform. Pain is placed between the two because it can occur because of both external and internal events.

A more complete discussion of this is in the Manual

INFLUENCE OF SENSORY LOSS ON DEVELOPMENT: The Communication Bubble

Are all five major senses working?

Estimates are quoted in the literature that the percentage of information that we take in via our eyes is anywhere from 60 to 90%. Of course, if you are listening to an audio tape, vision doesn't matter. If you are watching a film strip, hearing is of no consequence. In addition, some people are visual learners and some people are auditory learners. It is well-known that two people with identical audiograms may have strikingly different abilities to understand speech and other sounds. Perhaps the one who understands speech better is actually an auditory learner. But even that person misses information. If he/she uses speech-reading extensively, then a vision impairment on top of a hearing impairment will cut down on understanding as well.

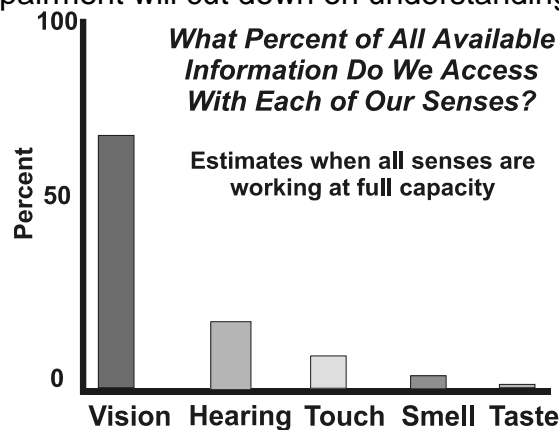
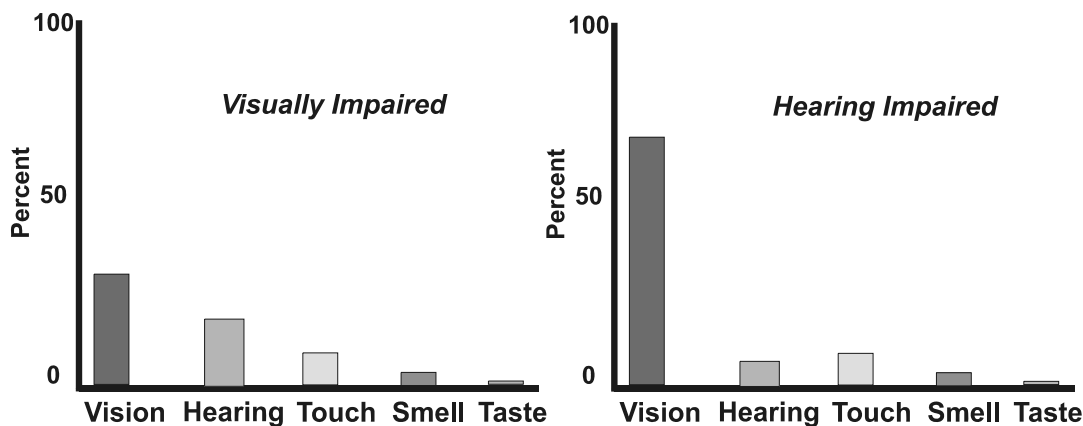


Figure 1: Full access to information from all senses.

However, consider what happens if a child is visually impaired AND hearing impaired. If a child has moderate visual impairment, the Vision bar might be half as high. If the child is moderately hard of hearing, the Hearing bar would be shortened by half. Relatively speaking, then, the other senses become more important.



Figures 2 & 3: Compare the differences when half of either vision or hearing is gone.

Now consider how less information is available when half of BOTH the vision and the hearing are missing. Notice how much more important the senses of touch and smell become.

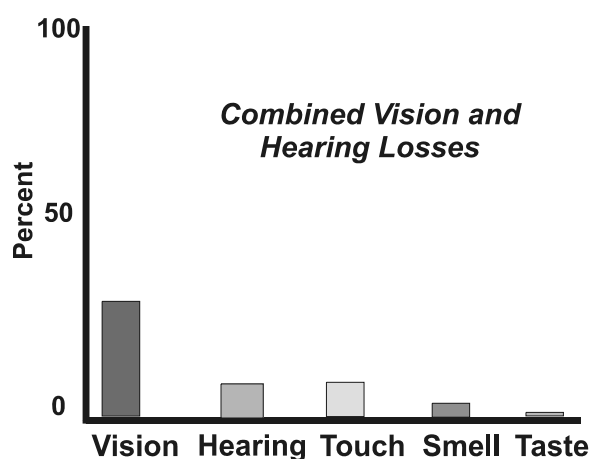


Figure 4: Combined vision/hearing loss with half of each gone.

What is Deaf-Blind?

Definition: Any combination of hearing and vision loss that interferes with access to communication and the environment and requires interventions beyond those necessary for hearing or vision loss alone. For educational purposes in most states, the child needs to meet the criteria for deaf/hard-of-hearing as well as for blind/visually impaired (check with your own DeafBlind Project director)

Deaf-Blind rarely means totally deaf and totally blind. No other succinct term has been found that fits all of the conditions listed in the table below. Dual sensory loss, Hard of Hearing/Visually Impaired, etc. are cumbersome. Most parents and educators have finally settled on DeafBlind even though that term is a somewhat inaccurate and, often, a scary term.

The importance of having a DeafBlind label, however, cannot be underestimated. The educational needs are truly different when both senses are affected. It is not enough to have the consultants come in and give their input without considering the effect of the other sensory loss. The reason is that the techniques used to compensate for vision loss often involve hearing and those used for hearing loss frequently involve vision. The combination makes a HUGE difference in the way information should be presented.

What combination of vision & hearing is present?

When one or more senses are impaired, additional educational consultants are needed, even (or perhaps especially) during the Early Childhood phase of development. Research out of Denver shows that, if a communication system is established BEFORE six months of age, a deaf child can develop completely normal language. Even if they learn to read sign language, they do not fall behind their hearing peers in expressive language and reading. The implications are staggering. Even though medical issues in CHARGE occupy the thoughts and minds of parents and caretakers during the first 2 years of life, it would appear to be very important to start some form of communication as early as possible.

The Need to Establish A Communication Bubble

Establishing what Susan Smith, the parent of two children with CHARGE, calls the “Communication Bubble” is essential if you want to make sure the child knows you are there and are trying to communicate. The concept of a “bubble” is a good one because you can imagine the space within which you need to be. As Eric Kloos says, you need to be “on the child’s radar.” Get outside the bubble and you might as well not exist. Too many times, we think the child is tuned out, not paying attention, or is too “retarded” to answer when, in fact, he/she may not even know that you are trying to say something.

Each parent should know how far a child can see what kinds and colors of objects, with or without movement, in different lighting conditions and against different shaded backgrounds. The eye doctor will usually not be able to tell you this except by guessing though some do this kind of testing. A vision teacher, however, can do a Functional Vision Evaluation, to help establish these visual abilities. Figuring out what the child hears clearly, at what distance and on which side is also important. Neither of these is easy to determine so parents and team members will likely have to engage in a continuing evaluation process.

The other very important point is that a lot of communication precedes formal language. Every mother can “read” her child’s communication regardless of whether that child uses any formal language. Sometimes it is guessing, but the child will let the mother know when she’s got it right. Therefore, it is important to lead up to formal communication step by step. We use sight cues and verbal cues like outstretched arms with some encouraging words to let a hearing/sighted child know what we want to do next. For a child without good vision or hearing, concrete object cues and touch cues are used instead. DeafBlind project staff can help the regular and special education staff learn how to use these methods. As parents and educators we may get hung up on wanting our children to speak our own language using the method we use. That is as natural as immigrants wanting their children to continue speaking their language and remembering their culture. However, the issue is not the form but the substance. The child needs to learn that an object, touch, picture, word or sign is a symbol that stands for a thought. Many of them strung together convey complex ideas or stories. They include naming of objects, actions, remembering the past, anticipating the future and, later, getting into abstract thoughts and discussions.

So, above all, just get to it! COMMUNICATE, COMMUNICATE, COMMUNICATE.



PROGRAM SECTION 17

Breakout Session #4
Friday, July 27, 2007, 4:15-5:30
Pacific Ballroom II

Psychiatric Diagnoses and
Psychotropic Medications: A
pediatric survey



Presented by
Lee Wachtel
and Tim Hartshorne

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

17 MG Psychiatric Diagnoses and Psychotropic Medications: A pediatric survey (Lee Wachtel and Time Hartshorne)

Many children diagnosed with CHARGE syndrome demonstrate behavioral difficulties in addition to visual, hearing and other systemic impairments. Previous research has reported that children with CHARGE have increased rates of self-injury and aggression, as well as increased frequency of obsessive compulsive and autism spectrum disorders. This study asked parents to report not only the diagnoses given for their child's behavior problems, but also whether psychotropic medication interventions were prescribed, and which agents were chosen. The results of this study showed that according to parental report, anxiety disorders and pervasive developmental disorders were the most common psychiatric diagnoses assigned, with antidepressant and antipsychotic medications the most frequently prescribed psychopharmacological agents.



PROGRAM SECTION 18

Breakout Session #4
Friday, July 27, 2007, 4:15-5:30
Pacific Ballroom II

Homeschooling Works:
Let the Journey Begin!



Presented by
Shirley Minster

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Benefits of Homeschooling

By Shirley M. R. Minster, M. S. Ed.

Home Education & Family Services

www.HomeEducator.com

PO Box 1056 / 51 West Gray Rd

Portland, ME 04039

(207) 657-2800

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

1

Benefits of Homeschooling for Children

- Curriculum tailored to the student
- Social skills develop naturally to be used with all ages
- Security in knowing that parents want to be with them
- Understand that an education is important
- Know how to work to understand new concepts and are rewarded with demonstration of the new knowledge
- Understand the teacher's teaching style because it remains consistent year to year
- Multitude of teaching styles in a day is not an issue
- Consistency in the teacher's values
- Can move on once concept is learned; no waiting period

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

2

Benefits of Homeschooling for Children, p 2

- Know that adults don't know everything and that's okay
- Learn how to search for answers
- Okay it's okay to get excited when learning
- Parents know what method is being used to teach concepts
- Children don't have to explain purpose of assignment
- Learning is more efficient
- Health is better
 - Inside when cold
 - Eat when hungry
 - Sleep when tired
 - Sit in sun
 - Exposure to germs lower
 - Laugh when happy
 - Use bathroom when necessary
 - Sit and think when want to

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

3

Beginning the Homeschool Journey

By Shirley M. R. Minster, M. S. Ed.

Home Education & Family Services /
Royal Academy

www.HomeEducator.com

PO Box 1056 / 51 West Gray Rd
Portland, ME 04039

(207) 657-2800

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Beginning the Homeschool Journey

Step1: Personal Preparation

- Read books by the Moores to get insight and facts about homeschooling. Be prepared!
- Visit our website for help:
www.HomeEducator.com
- Read *Home Educator's Family Times* for up-to-date materials, experienced homeschoolers' tips, and encouragement.

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Beginning the Homeschool Journey

Step 2a: Talk with Others

- Legalities in your state and/or city
- Available resources
- Homeschool support groups
- Classes for the children
- Curriculum they use
- Field trips
- Conferences

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Beginning the Homeschool Journey

Step 2b: Talk with Others

- Typical yearly cost
- Involvement in local public or private school
- Work options for teens
- How they filled out paperwork, if needed, to homeschool

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Beginning the Homeschool Journey

Step 3: Talk with Your Children

- Their concerns
- Topics they want to study
- Interests
- Difficulties they have
- Study area needs

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Beginning the Homeschool Journey

Step 4: Setting Up the Homeschool

- Decide where most of schooling will be done.
- Get ample supplies (pencils, paper, markers, etc.).
- Consider tables, shelving, and lighting.
- Purchase academic supplies.
- Consider having individual standardized achievement tests done for each child.

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Choosing Curriculum by Shirley M. R. Minster, M. S. Ed.

Think Before You Buy!

By taking the time to investigate either early in the school year or at the end of the year while preparing for the new year, you will save time and money. You will not end up trying to work with a curriculum is not a good.

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Choosing Curriculum

- I like this curriculum because...
- Its strengths are...
- One area that makes this curriculum difficult to use is:
- Its weaknesses are
- Extra materials needed (science equipment, construction paper, audio-visual extras, charts, etc.)?
- Cost to buy the extra materials or are they included in a package deal?
- Enough supplemental materials and activities to stimulate a child who has special needs or is gifted?
- Overall cost of the curriculum?

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Choosing Curriculum

- Possible to buy individual pieces as needed or must purchase a whole package?
- Do the children like the curriculum? Why / Why not?
- Can the teacher be flexible or is there just one way to use it?
- Are the materials consumable (workbooks)?
- Is a teacher's book available? Is it a teacher's guide with lesson plans, activities, suggestions or simply an answer key?
- Are pictures and illustrations colorful or black-and-white sketches?

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

Choosing Curriculum

- Are graphs, illustrations, and written examples clear or confusing?
- Are pictures helpful or distracting?
- Is the student book 'user friendly'? Does it inspire success or does it seem overwhelming in its approach?
- Is the font size appropriate for the age of the child?
- What is your child's learning style? Does the curriculum fit this style?

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

The Homeschool Schedule

By Shirley M. R. Minster, M. S. Ed.

Home Education & Family Services /
Royal Academy
www.HomeEducator.com
PO Box 1056 / 51 West Gray Rd
Portland, ME 04039
(207) 657-2800

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

1

The Homeschool Schedule

- Organize the home for efficiency of time
- Consider the ages and stages of children
- Plan for changes in the family
- Make a schedule that fits everyone's needs
- Change in chore assignments to fit child
- Decide which academic courses work best in what part of the day
- Decide on choice of monthly or seasonal studies

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

2

The Homeschool Schedule

- Schedule vacation time
- Schedule 'down time' for tutors
- Include children's personal interests
- Plan activities for individual children as well as family as a group
- Resist temptation to add too many outside activities (sports teams, private lessons, clubs, etc.)

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

3

The Homeschool Schedule

- Remember the littlest ones need their special time, too, but to not infringe on older children's study time.
- Plan in break times (snacks, rest, outdoors, etc.)
- Limit computer time and television time.
- Plan in arts and crafts.
- Plan in quiet time for everyone.

7/17/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

4




Homeschooling Children with Special Needs

Shirley M. R. Minster, M. S. Ed.
Home Education & Family Services
www.HomeEducator.com

7/18/2007

Shirley M. R. Minster, M. S. Ed.
© 2007




My Advice to Parents

- 1. Be practical.
- 2. Be consistent.
- 3. Be firmly gentle.
- 4. Know your child's rights.
- 5. Keep a sense of humor.
- 6. Keep a daily schedule ... for sanity's sake.
- 7. Get into your corner, pray, reflect.

7/18/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

- 
- 8. Pick your support team carefully (family member(s), friend(s), other care givers, etc.).
 - 9. Read books about the special need.
 - 10. Go to conferences.
 - 11. Visit internet sites for encouragement, help, and updated information.
 - 12. Think about the future as well as the present.

7/18/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

And Finally ...



- 13. Realize that the present really is a present. Your child is a special gift to you.

7/18/2007

Shirley M. R. Minster, M. S. Ed.
© 2007

A Bibliography for Homeschooling and Special Needs

Shirley M. R. Minster, M. S. Ed.

www.HomeEducator.com

- Adee, Donna and Ellis Adee. God's Special Child: Lessons from Nathan and Other Children with Special Needs. Harvest Publications, KS: 1996.
- Alexander-Roberts, Colleen. ADHD & Teens: A Parent's Guide to Making It Through the Tough Years. Taylor Publishing Company, Texas: 1995.
- Amen, Daniel G. Healing ADD: The Breakthrough Program That Allows You to See and Heal the 6 Types of ADD. G.P. Putnam's Sons, New York: 2001.
- *He has also written other books on this topic: A Teenager's Guide to ADD, A Child's Guide to ADD, A Sibling's Guide to ADD, A Teacher's Guide to ADD.
- Andreola, Karen. A Charlotte Mason Companion: Personal Reflections on the Gentle Art of Learning. Charlotte Mason Research & Supply: 1998.
- Attwood, Tony. Asperger's Syndrome: A Guide for Parents and Professionals. Jessica Kingsley Publishers Ltd, PA: 1998.
- Barnier, Carol. How to Get Your Child Off the Refrigerator and on to Learning. Emerald Books, Washington: 2000.
- Blakemore-Brown, Lisa. Reweaving the Autistic Tapestry: Autism, Asperger Syndrome and ADHD. Jessica Kingsley Publishers Ltd, PA: 2002.
- Citro, Teresa Allissa, Editor. Transitional Skills for Post Secondary Success ... Reflections for Students with Learning Disabilities. Learning Disabilities Association of Massachusetts: 2001.
- Coleman, Jeanine B. The Early Intervention Dictionary. Woodbine House, , MD: 1999.
- Davis, Ronald D. The Gift of Dyslexia. Ability Workshop Press, CA: 1994.
- Dwyer, Kathleen. What Do You Mean I Have a Learning Disability? Walker and Company, New York: 1991.
- Freed, Jeffrey and Laurie Parsons. Right-Brained Children in a Left-Brained World: Unlocking the Potential of Your ADD Child. Fireside, 1998.
- Hartmann, Thom. Attention Deficit Disorder: A New Perception. Underwood Books, 1997.
- Healty, Jane M. Your Child's Growing Mind. Doubleday, NY: 1989.
- Hensley, Sharon C. Home Schooling Children with Special Needs. Noble, WA: 2001.
- Hoekstra, Elizabeth and Mary Bradford. Chronic Kids, Constant Hope. Crossway Books, Wheaton, IL: 2000.
- Hollowell, Edward M. and John J. Ratey. Driven to Distraction: Recognizing and Coping With Attention Deficit Disorder from Childhood Through Adulthood. Simon & Schuster, 1995.
- Hollowell, Edward M. and John J. Ratey. Answers to Distraction. Bantam Books, 1996.
- Hyatt-Foley, DeAnn and Matthew G. Foley. Getting Services for Your Child on the Autism Spectrum. Jessical Kingsley Publishers, PA: 2002.
- Kelly, Kate and Peggy Ramundo. You Mean I'm Not Lazy, Stupid or Crazy?! : A Self-Help Book for Adults With Attention Deficit Disorder. Fireside, 1996.
- Kranowitz, Carol Stock. The Out-of-Sync Child Has Fun: Activities for Kids with Sensory Integration Dysfunction. The Berkley Publishing Group, NY: 2003.
- Kravets, Marybeth and Imy F. Wax. The K & W Guide to Colleges for the Learning Disabled. HarperPerennial, NY: 1993.
- Martin, Katherine. L. Does My Child Have a Speech Problem? Chicago Review Press, IL: 1997.
- McAfee, Jeanette, M. D. Navigating the Social World: A Curriculum for Individuals with Asperger's Syndrome, High Functioning Autism and Related Disorders. Future Horizons, Inc., Texas: 2002.
- Moore, Raymond S., Dennis R. Moore, and Dorothy N. Moore. Better Late Than Early: A New Approach to Your Child's Education. Reader's Digest Association, 1989.
- Moore, Dorothy N. and Raymond S. Moore. The Successful Homeschool Family Handbook. Thomas Nelson: 1994.

A Bibliography for Homeschooling and Special Needs, p 2

- Moore, Raymond S. and Dorothy N. Moore. School Can Wait II. Brigham Young University Press: 1989.
- Murphy, Kevin R. and Suzanne Levert. Out of the Fog : Treatment Options and Coping Strategies for Adult Attention Deficit Disorder. Hyperion, 1995.
- Nadeau, Kathleen G. Adventures in Fast Forward : Life, Love, and Work for the ADD Adult. Brunner-Mazel Trade, 1996.
- Parker, Harvey C. Put Yourself in Their Shoes: Understanding Teenagers with Attention Deficit Hyperactivity Disorder. Specialty Press, Inc, Florida: 1999.
- Saunders, Jacquelyn with Pamela Espeland. Bringing Out the Best: A Resource Guide for Parents of Young Gifted Children. Free Spirit Publishing Inc.. MN: 1991.
- Sears, William and Lynda Thompson. The A.D.D. Book. Little, Brown, Boston: 1998.
- Snelick, Richard and Karla Dougherty. Living with Brain Injury: A Guide for Families. Healthsouth Press, AL: 2001.
- Solden, Sari. Women with Attention Deficit Disorder. Underwood Books, 1995.
- Stevens, Suzanne H. The LD Child and the ADHD Child: Ways Parents and Professionals Can Help. John F. Blair, Publisher, Winston-Salem, NC: 1996.
- Stordy, B. Jacqueline, Ph.D. and Malcolm J. Nicholl. The LCP Solution: The Remarkable Nutritional Treatment for ADHD, Dyslexia and Dyspraxia. Ballantine Books, New York: 2000.
- Van't Land, Marcia. Living Well with Chronic Illness. Harold Shaw Publishers, IL: 1994.
- Walker, Sally Yahnke. The Survival Guide for Parents of Gifted Kids. Free Spirit Publishing, Minneapolis: 1991.
- Weiss, Lynn. Attention Deficit Disorder in Adults. Taylor Pub, 1997.
- Wender, Paul. ADHD: Attention-Deficit Hyperactivity Disorder in Children and Adults. Oxford University Press, 2000.
- Wender, Paul. The Hyperactive Child, Adolescence, and Adult: Attention Deficit Disorder Through the Lifespan. Oxford University Press, 1987.
- Wood, Chip. Yardsticks: Children in the Classroom Ages 4-14. Northeast Foundation for Children, MA: 1997.
- Wright, Pam and Pete Wright. From Emotions to Advocacy. Harbor House Law Press, Virginia: 2004.
- Zelinsky, Deborah. The Mind-Eye Connection: Open Your Eyes and Listen. The Mind-Eye Connection, P.C.: 2002.
- Zimmerman, Marcia. The ADD Nutrition Solution : A Drug-Free Thirty-Day Plan. Owl Books, 1999.

Websites to Visit – Due to website addresses changing, caution is advised.

Home Education & Family Services/Royal Academy Online Bookstore: www.homeeducator.com

Adult ADD/ADHD Resource Guide: <http://www.addresources.com>

Born to Explore! The Other Side of ADD by Teresa Gallagher: <http://borntoexplore.org/>

CHADD (Children and Adults with Attention-Deficit/Hyperactivity Disorder): <http://chadd.org>

Magazines / Journals

Home Educator's Family Times: www.HomeEducator.com/FamilyTimes

PO Box 6442, Brunswick, ME 04011

NATHHAN News (National Challenged Homeschoolers Associated Network): www.nathhan.com

PO Box 310, Moyie Springs, ID 83845, 208-267-6246



PROGRAM SECTION 19

Breakout Session #4
Friday, July 27, 2007, 4:15-5:30
Pacific Ballroom III

Transition-to-Adulthood:
Getting Connected



Presented by
Susan Ruzenski, Philip
Wisner, & Cathy Kirscher

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

HELEN KELLER NATIONAL CENTER VOCATIONAL SERVICES

EMPLOYMENT

**Appreciation of
Life Experience**

**Career
Exploration &
Counseling**

Job Development

Work Experience
(on- the- job training)

OUTCOME

CONCEPTUAL FRAMEWORK

- Person Centered
- Consumer Determined
- Measurable Goals
- Transdisciplinary Training
- Collaborative Teamwork

The Vocational Services Department at HKNC is committed to empowering individuals who are deaf-blind to develop skills and resources in order to obtain gainful employment that is commensurate with their aspirations and abilities. Over the last five years the employment outcomes of consumers attending HKNC has ranged between 38-54%.

The Department's overall mission is to support each consumer in obtaining meaningful employment in their home community by working collaboratively with the consumer, the state Vocational Rehabilitation Counselor and local employment agencies.

Vocational Evaluation and Training Resulting in Employment

This program is designed to assist the consumer in choosing feasible career goals by developing work interests and skills.

Services Framework:

✓ **Appreciation of Life Experience** - self awareness of preferences and strengths. Personal Futures Planning is a process to guide consumers through a realization of their interests and experiences and links those to vocational goals.

✓ **Formal and informal assessment** through testing, situational assessments or informational interviews to determine:

Interests, Aptitudes
Academic skills, Preferences
Instructional Procedures

In preparation for employment, consumers have an opportunity to increase productivity and work stamina, identify job modification and accommodations, develop communication strategies with employers and co-workers and travel independently to and from work.

Group discussion, peer collaborative learning and work adjustment group provides a forum on challenges faced during experiences and a sharing of expectations and perceptions.

Additional activities include: resume preparation, job searching via the internet, communication with co-workers and supervisors, interviewing skills, information on company benefits and social security programs as well as a discussion of the five titles of the Americans with Disabilities Act.

Development of Employment Plan -The HKNC Employment Plan is developed by the consumer with assistance from the Employment Training Specialist and other members of the team. The Employment Plan states the consumer's intended vocational outcome and identifies the skills the consumer must master to obtain the outcome. Each skill area includes information regarding the industry's standards for employment and the consumer's progress towards meeting these standards. Progress towards the vocational outcome is monitored by the consumer and the Employment Training Specialist with the consumer determining the ways in which progress will be assessed.

Job Development- Early in the evaluation, information about job opportunities in the home community is pursued. The key to successful vocational outcome is working closely with the team in the home community. With the collaboration from state VR, HKNC affiliates/Regional Representatives and the community providers, resources and support needs are identified and a transition plan is initiated. Consumers play a primary role in the process of developing an

employment-based transition plan which is continually modified during the entire training process.

The consumer, with support from the transdisciplinary team, identifies job opportunities available in the home community. Job interviews are scheduled and visits to the consumer's home community may occur during the final training period at the Center or immediately upon completion of the program. Consumers develop a resource book and are encouraged to contact local resources for any additional support.

Currently, the vocational training program has affiliations with several nationally known companies where job opportunities may become available. These include:

<i>1-800-Flowers</i>	<i>Gap</i>
<i>TJ Maxx</i>	<i>Auto Zone</i>
<i>Tower Records</i>	<i>Wal-Mart</i>
<i>Filene's Basement</i>	<i>Tower Records</i>
<i>Social Security Office</i>	<i>Petco</i>
<i>McDonald's</i>	<i>Burger King</i>
<i>Big Brothers/Big Sisters</i>	<i>Office Max</i>
<i>Auto Barn</i>	<i>Waldbaums</i>
<i>YMCA</i>	<i>Sears</i>
<i>Michael's Crafts Store</i>	<i>Tutor Time</i>
<i>Home Depot</i>	<i>JP Morgan Chase</i>
<i>Luxottica</i>	<i>Ametek</i>
<i>Thermal Systems</i>	

Work experience positions include but are not limited to: office clerk, animal groomer, stock associate, bookstore clerk, library page, computer repair associate, assembler/packager for a manufacturer of adaptive equipment, data entry clerk, technology tutor, accounts payable clerk, adaptive technology instructor, customer services representative, disc jockey assistant and presenter in a children's museum. Local work sites include: St. Francis Hospital, Sunny Marketing, Port Washington Library, Minute Man Press, Stop and Shop, Alper's Hardware, Forest Value Books, Compustar USA, Great Neck Art Center, Jo Ann's Fabrics, Five J's Jewelers, Manhasset Animal Hospital, DeMatteis Center, Independent Living Aids, Nassau Community College, Sunrise Mall, Country Dry Cleaners, Gulfway Marine Services, Clever Devices, Sands Point Nursing Home, Let's Wrap, Central Credit Union, LDI (Leslie Digital Imaging), Pulse Magazine, Cleary School for the Deaf, Automotive Unlimited and AHRC.

Commenting on her vocational training at HKNC, a former consumer stated: *"I realized ...that without a doubt this is the fulfillment of my lifelong dream. This training opportunity will have such a positive impact on my life and help me to reach my vocational goal."*

For further information, please contact:
Admissions Coordinator
(516) 944-8900 (voice)
(516) 944-8637 (TTY)
or your Regional Representative

HELEN KELLER NATIONAL CENTER

Telephone: 516-944-8900

FAX: 516-944-7302

SUMMER EVALUATION PROGRAM

Make it Happen! It's up to YOU!

Come join us at HKNC this summer! Are you 16 years of age or older? Currently attending high school? Would you like to participate in career exploration or in identifying your next steps to college?

HKNC's transdisciplinary program offers individualized evaluation services, in a variety of areas. This program is designed to offer you information and practical experience to expand your awareness, knowledge and skill in the following areas:

Vocational Exploration

Accessing College and Community Supports and Services

Adaptive Technology and other Communication Aids and Devices

Skills of Independent Living

Self- Determination and Advocacy

Communication Methods

Interpreting Issues

Orientation and Mobility

Low Vision

Audiology

Participating in leisure and recreational activities during evenings and weekends while meeting new friends is also part of the HKNC Summer Evaluation program experience. Meet and learn from others who are successful!

GOAL/OUTCOME: Participate in a variety of training experiences in order to:

- ☆ gain information and resources
- ☆ network with others
- ☆ assess your strengths and weaknesses
- ☆ determine future training objectives
- ☆ meet new friends

You will have an opportunity to develop a personalized plan that will support you in reaching your goals after high school graduation.

NUMBER OF PARTICIPANTS: 8 - 10

LENGTH OF TRAINING: Evaluation is typically 8 weeks.

For further information, please contact:

Admissions Coordinator

(516) 944-8900 ext 258 (voice/relay)

Helen Keller National Center
141 Middle Neck Road, Sands Point, NY 11050
516-944-8900 (Voice)/ 516-944-8637 (TTY)
516-944-7302 (fax)
hkncinfo@hknc.org
www.hknc.org

Person Centered Approach Toward Habilitation (PATH)

The PATH program offers evaluation and training services to consumers who are deaf-blind and developmentally disabled. Services reflect individual needs, preferences, abilities and interests in the domains of work, home and the community. Each participant in the PATH Program has an opportunity to gain desirable skills for integrated employment and community living.

Communication

The diverse and unique communication preference of each participant is fostered through meaningful dialogue while striving toward a more formal conventional symbol system or language. Communication methods preferred by consumers include: object symbols, sequence boxes, communication cards, gestures, sign language, pictures and written language. Natural, spontaneous, ongoing conversations give communication meaning.

Orientation and Mobility Training

The PATH program offers evaluation and training services to consumers who are deaf-blind and developmentally disabled. Services reflect individual needs, preferences, abilities and interests in the domains of work, home and the community. Each participant in the PATH Program has an opportunity to gain desirable skills for integrated employment and community living.

Vocational Assessment

The vocational interests, abilities, training and support needs of each consumer are assessed in integrated community settings, through the use of trial work experiences with appropriate supports. It is our belief that any individual is “presumed” able to benefit from the opportunity to explore an employment outcome.

Through a variety of work experiences, a consumer has an opportunity to demonstrate increased independence and that he/she can benefit from continued training to achieve a vocational outcome with appropriate support. Work experiences are conducted over a period of time which allows for personal adjustment while also strengthening job performance, learning appropriate work behaviors and increasing work tolerance. Through consistent communication, and individualized instruction, carried out by qualified Helen Keller National Center personnel, consumers build a foundation of skills opening the pathway to employment.

Vocational Training

On-the-job training occurs in the community at a variety of work sites such as a local hospital, supermarket, nursing home, retail and craft stores, horse stable, restaurant, farm and manufacturing company. Communication skills, orientation and mobility strategies and adaptive technologies are also used as the student works to master specific work skills.

Community Living Skills

Consumers gain experience and skills in preparation for successful participation in group and supported living in their home community. Training and support are provided by a core group of staff in an on-campus apartment setting focusing on all natural activities of adult life.

Transition and Futures Planning

Transition planning toward community employment and life enrichment is conducted through a collaborative effort of the transdisciplinary staff of the Helen Keller National Center (including the dedicated PATH staff), family members, community members, vocational rehabilitation counselors, affiliated agencies and other community service providers.

The Helen Keller National Center is integrally involved with the process of finding a home for the consumer that matches his/her individual preferred lifestyle. HKNC offers training to community service providers and the consumer's family members in order to successfully meet the lifestyle, communication, behavioral and community living preferences of the consumer.

For more information about the PATH Program, or any of the services provided by the Helen Keller National Center, call Information Services at 516-944-8900 x 326 or e-mail at hkncinfo@hknc.org.



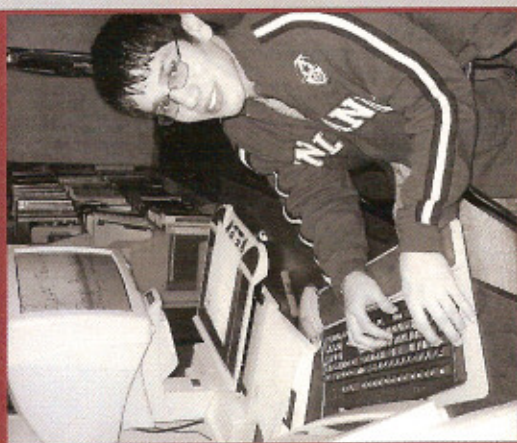
Helen Keller National Center
For Deaf-Blind Youths and Adults

141 Middle Neck Rd.
Sands Point, NY 11050



Helen Keller National Center
For Deaf-Blind Youths and Adults

Programs & Services



141 Middle Neck Rd.
Sands Point, NY 11050

Phone: (516) 944-8900

TTY: (516) 944-8637

Fax: (516) 944-7302

E-mail: hkncinfo@hknc.org

Web: www.hknc.org

Regional Representatives

Staff in the regional offices offer the following services, free of charge, to agencies and to individuals who are deaf-blind:

- Consumer advocacy
- Consultation and technical assistance to schools and agencies
- Assistance in developing local services
- Consumer follow-along support "as needed"
- Information and referral
- Professional development and in-service training
- Public education and awareness
- Maintenance of information for National Registry

Supervisor of Regional Representatives:

Susan Lascek
1003 Virginia Ave., Suite 106
Atlanta, GA 30354
(404) 766-5800 (V/TTY)
(404) 766-3447 (FAX)
SLHKNC4@aol.com

I. New England Region
CT ME MA NH RI VT
Mary Ellen Barbiasz
(781) 259-7100 (V/TTY)
HKNC1meb@comcast.net

II. Mid-Atlantic Region
NJ NY PR VI
Nancy Godfrey
(516) 944-8900 Ext 293(V)
(516) 883-6914 (TTY)
HKGodfrey@aol.com

III. East Central Region
DE DC MD PA VA WV
Cynthia L. Ingraham
(301) 459-5474 (V)
(301) 459-5433 (TTY)
HKNCREG3CL@aol.com

IV. Southeastern Region
AL FL GA MS
Amy Parker
ap@hknc.org
KY NC SC TN
Monika McJannet-Werner
MW4HKNC@aol.com
(404) 766-9625 (V)
(404) 766-2820 (TTY)

V. North Central Region
IL IN OH MI MN WI
Laura J. Thomas
(309) 755-0018 (V/TTY/VP)
(309) 755-0021 (V/TTY)
HKNC5LT@aol.com

VI. South Central Region
AR LA NM OK TX
CC Davis
(972) 490-9677 (V/TTY/VP)
CCFLUTBOL@aol.com

VII. Great Plains Region
IA KS MO NE
Beth Jordan
(913) 677-4562 (V/TTY)
hknc7BJ@aol.com

VIII. Rocky Mountain Region
CO MT ND SD UT WY
Maureen McGowan
(303) 934-9037 (V/TTY)
Maureen.McGowan@hknc.org

IX. Southwestern Region
AZ CA HI NV, Guam, Samoa,
Commonwealth of Northern
Mariana Islands
Cathy Kirscher
(858) 623-2777 Ext 389 (V)
(858) 646-0784 (TTY)
CKirscher@alliant.edu

X. Northwestern Region
AK ID OR WA
Dorothy Walt
(206) 324-9120 (V)
(206) 324-1133 (TTY/VP)
NWHKNC@juno.com



Programs & Services

Vocational Training Program

Headquartered in Sands Point, NY, the HKNC training program provides adults who are deaf-blind:

- Evaluation
- Short-term comprehensive vocational rehabilitation training
- Assistance in obtaining employment, housing and community supports

Recognizing the unique talents, strengths and desires of each consumer, the Vocational Training Program is tailored to provide learning opportunities and choices which parallel each person's future lifestyle at home, work and in the community.



Additional programs offered at Headquarters:

- Two-week summer seminar for high school students
- Eight-week summer evaluation and training for high school students
- Two-week summer seminar for senior adults
- Professional Internships

For more information, contact your Regional Representative.



Affiliate Program

HKNC provides financial assistance to more than 40 state and private agencies to serve people with deaf-blindness and enhance local service capability. New grants are awarded on a competitive basis.

Contact: Nancy Godfrey - Coordinator;
(516) 944-8900 Ext. 293 (Voice);
(516) 883-6914 (TTY);
E-mail: HKGodfrey@aol.com

Senior Adult Program

HKNC's senior adult specialist provides:

- Consultation
- Training
- Technical assistance

These services are available to families, consumers and professionals nationwide to help them to develop a greater understanding of the abilities and needs of senior adults, age 55 and up, who are deaf-blind or who are experiencing age-related vision and hearing loss. The specialist networks with local, state, and national organizations that serve senior adults.

Contact: Paige Berry - Senior Adult Specialist;
(804) 827-0920 (Voice/TTY);
E-mail: hkncpaigeap@aol.com

National Training Team (NTT)

The National Training Team conducts seminars at headquarters. They provide training on a variety of topics related to deaf-blindness, such as:

- Introduction to Deaf-Blindness
- Communication
- Employment Training
- Orientation & Mobility
- Family Issues
- The Elderly Population
- Assistive Technology
- Person Centered Planning & Transition

The NTT also provides on-site training for agencies or organizations nationwide. Each program is tailored to meet specific local needs.

Contact: Sr. Bernadette Wynne - Coordinator;
(516) 944-8900 Ext. 239 (Voice);
(516) 944-8637 (TTY);
E-mail: NTTHKNC@aol.com

National Family Association For Deaf-Blind (NFADB)



NFADB is an independent, not-for-profit national family organization, partially funded by HKNC. NFADB's goals include advocacy for all persons who are deaf-blind, sharing information and resources, facilitating family/professional partnerships and supporting national policies to benefit people with deaf-blindness. NFADB publishes a newsletter and has paid memberships.

Contact: (800) 255-0411;
E-mail: NFADB@aol.com;
Website: www.nfadb.org

HELEN KELLER NATIONAL CENTER FOR DEAF-BLIND YOUTHS & ADULTS

141 Middle Neck Rd.
Sands Point, NY 11050
Phone: (516) 944-8900
TTY: (516) 944-8637
Fax: (516) 944-7302
E-mail: hkncinfo@hknc.org
Web: www.hknc.org



PROGRAM SECTION 20

Breakout Session #4
Friday, July 27, 2007, 4:15-5:30
Pacific Ballroom IV

CHARGE 103 for New Families

Communication
and
Transition to School



Presented by
Rob Last

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

CHARGE 103: Babies Can't Wait To Communicate

The importance of communication in the early years and getting started early

Presentation by Rob Last
8th International CHARGE SYNDROME Conference
Costa Mesa, California USA
July 27 – 29 2007

Introduction

In the first few months of life children who are born with CHARGE Syndrome face numerous serious medical interventions. These interventions are often life saving and essential to ensure the child's long-term survival.

We know the children access a large range of medical specialists, therapists and educators.

We know that medical procedures mostly decrease as the children grow and develop.

We know the educational options are diverse ranging from placement in a specialist school setting through to integration and inclusion in a regular school.

We know the broad range of outcomes for these children from dependent to independent.

We know the broad range of communication systems used by the children.

We also know of these children's iron will and captivating personalities.

In this article I want to highlight the importance of communication - communication from the very beginning. The title, *Babies Can't Wait To Communicate*, alludes to children's early communication; the communication parents learn to understand so quickly.

- the cry at birth, the weak cry, the robust cry, the absent cry
- the cry to say; 'I'm hungry'
- the cry to say; 'I'm distressed' followed by contentment when comforted
- the cry to say; 'that hurts'
- the peaceful expression to say; 'I'm content'
- the expression, which says; 'I like being held by you'
- that first smile in return to a smile

All of those early communication attempts which take place between babies and parents.

So What is Early Communication?

Early communication is an expression of a need or a reaction to a situation, which is recognised and responded to by another individual.

Why the Need to be Aware of Early Communication?

The child who has CHARGE Syndrome may have a vision loss, a hearing loss, reduced sense of smell, reduced sense of taste and a sense of touch that may not be as sensitive. Therefore all five senses may be affected.

In addition the child may also have additional challenges in breathing, managing excessive secretions, facial palsy and in some cases a cleft palate.

So it's suggested to incorporate a range of communication strategies to provide every opportunity for the child to make sense of their already confusing world.

Early communication strategies in the hospital setting

- Position the hospital crib so the baby's vision isn't compromised further by light from a window or overhead light, ensuring the subject of interest for the baby is well lit. These may be the faces of the parents, brothers and sisters, the interesting toy, the music box, the shiny beads or the colourful mobile hanging overhead.
- Alert the baby through a simple motor cue that something is about to occur. A tummy rub to indicate bath time, pressure on the hand prior to a needle being inserted, a gentle upward lifting motion of the baby's body prior to being lifted up.
- Sing songs, paired with gentle movements, close to the baby's ear.
- Provide a pat, pat, pat, on the child's back prior to physiotherapy.

Each of these simple actions provides an opportunity for the baby to understand what is about to happen and to anticipate that event, no matter whether it's a pleasant or unpleasant one.

By incorporating a motor movement to indicate what is about to happen we also provide an opportunity for the baby to use a motor movement to communicate a need.

It's been my experience these strategies may not be incorporated into family life until the baby comes home from hospital, I would encourage these strategies be incorporated into the baby's life from as early as possible, particularly in the hospital setting.

Speech and Communication

Speech is just one of the forms in which we communicate. We also use body language, facial expression, natural gestures, situational context, in fact all senses to gain additional information. Just consider bath time; the sound of the water running, the brightness of the room, the smell of the soap, the touch of the towel, the taste of the water.

So to incorporate a holistic approach to communication and to dispel the idea of speech being the only form of communication is thought to be appropriate in the overall communication needs of a child with CHARGE.

As already mentioned, the early need to alert the child to the events occurring to them in their environment is critical. At the same time gathering information on the various communication options and what is involved in adopting these options is also critical.

Questions families may want to ask in seeking information on the various communication options:

- **Sign language**

What is sign language? Who uses sign language?
How is it learnt? Is it difficult to learn? Which family members need to learn it?
How does my child get educated using sign language?
Who will understand our child other than family members?
Can we meet someone who uses sign language?

- **Speech using audition through hearing aids**

What is the degree of deafness in each ear?
How early can hearing aids be fitted? How will we know speech is being heard?
What can we expect our child's speech to be like?
What is the impact of facial palsy, breathing difficulties, excessive secretions and cleft palate on my child's ability to acquire speech?
Can we meet a family who has a child who has hearing aids and uses speech?

- **Speech using audition through a cochlear implant**

What is a cochlear implant? How and where is it fitted?
What kind of sound signal does it provide?
Does my child meet the criteria for cochlear implantation?
Are my child's cochleas intact enough to have an implant?
Can my child learn to speak using a cochlear implant?
How successful have cochlear implants been for other children with CHARGE?
Can we meet a family who have a child who has a cochlear implant?

- **Visual programs to support communication**

Real objects, object symbols, pictures of real objects, symbolic pictures, programs such as 'Board Maker' and 'Compic'. What are these? How do they work? Can my child see them?

Can these systems be of help in developing complex language?

Can you show us examples of what these systems look like and explain how they work?

- **Signed English**

Why don't we sign what we say in English?

Why is sign language, such as American Sign Language different from English? How will my child acquire English if using a sign language system such as American Sign Language?

Why are there two sign language systems?

Which signing system is better for our child and us?

- **Written English**

What does reading and writing have to do with communication?

How do we introduce books to our child? When do we introduce books to our child?

Can our child see the pictures and the text? How will my child learn to read?

What are the stepping-stones to literacy?

If my child acquires American Sign Language as their first language how will they access written English?

These questions are important to ask of the various relevant professionals and the answers provided should be factual and unbiased.

What Then is the Strategy?

From all this overwhelming complex information, perhaps the answer is simpler than it may seem.

My view is to incorporate a range of communication systems into family life and therefore immerse the child into a world of meaningful communication.

COMMUNICATION THAT IS MEANINGFUL TO YOUR CHILD

We already have lots of natural communication skills: body language, facial expression, natural gesture, as well as speech, which forms part of our everyday communication. What I believe we then do is layer informal and formal visual communication systems on top. For example; sign language, photos, drawings, real objects, symbolic objects, picture books, the written word, excellent hearing aids or a cochlear implant and quality FM listening systems.

How Does This Work?

With the early communication considerations in place, for example the cues and prompts to alert the baby to events, we also use speech to accompany these prompts and cues. We then add gestures to provide more meaning and also to provide the child with a means of expression. We still continue to talk.

We commence learning American Sign Language or a signing system applicable to the situation. Hearing aids are fitted as early as possible. We consider all the questions and issues surrounding a cochlear implant. Sign language skills are developing; therefore a more complex visual language is developing. We still continue to talk.

We continue to provide and maintain the best listening devices available. Books have been part of the child's life since the early months. Photos, drawings, video and audio tape can be used to create personalized stories and books. We are still talking.

Visual language skills have developed even further. By now the child and the family may be well on the way to achieving a communication system, which is meaningful to the child. The proof of this will be in the child's responses.

If the family decides to proceed with a cochlear implant, the process is well supported by visual language; the assessments, the preparation for surgery, the surgery, the technology, the switch on and the therapy.

Auditory and visual language can and should co-exist.

Communication and Behavior

If a child does not develop a communication system whereby feelings, thoughts, ideas, opinions and emotions can be expressed, they may be at risk in developing challenging behaviors; behaviors that are highly communicative yet expressed in a challenging manner. In order to minimize the risk of these behaviors and to manage them through communication, if they do arise, then the efforts in providing a meaningful language will be more than rewarded.

What Then is the Aim?

- To provide children with a range of communications.
- To ensure the communication systems are meaningful to the child.
- To ensure the communication provides opportunities to express feelings, emotions, thoughts and ideas in a rich and complex communicative environment.
- For the child to be immersed in a communication system which is meaningful at home, at preschool and at school.

Children's Stories

- Ella was provided with the opportunity to be bilingual, i.e. speech and Australian Sign Language. Over time her preferred first language became spoken English, however Ella chooses to use Australian Sign Language when experiencing breathing difficulties or auditory fatigue.

- Sam has good aided hearing in his right ear and is able to understand much of what is said in good listening conditions, but has difficulty speaking clearly due to his facial palsy. Australian Sign Language is his preferred means of communication.
- Brent has a cochlear implant and has acquired speech but also relies on sign language to assist in his understanding when the listening environment is poor or when he is tired and needs a rest from listening.
- Talea is a teenager who communicates fluently using Australian Sign Language. A while ago she developed some behaviors, which were of concern to her family. One being she would pick at her finger till it bled and another being where she would click her tongue loudly, a habit which annoyed her family. These were managed by asking Talea to wear gloves when she would feel like picking her finger and to chew gum when she would feel a need to click her tongue. These are language based interventions. Her family believe these behaviors may have escalated if Talea's communication system was at a level whereby they were not able to discuss these issues with her.

Children With CHARGE Seven Years Later – The Video

In 1993, at the first international conference for children with CHARGE in St Louis, Missouri, I presented a video of children with CHARGE using a range of communication systems. In 2000 I revisited some of these children to record their progress.

My main motivation in doing the follow up was because the early CHARGE literature spoke of all children with CHARGE having an intellectual disability, the R in CHARGE being 'retarded growth development and intellectual retardation'.

From the children I knew in Australia and many of the children I had met at the St Louis, Portland, Boston, Houston, Indianapolis, Cleveland and Miami conferences, it was clear to me that the children may have developmental delays due to their sensory disabilities but I felt many did not have an intellectual disability.

The outcomes in the video depict a broad range of abilities. Parents have reported viewing the video can be challenging but ultimately helpful. The video is available through the CHARGE SYNDROME Association – Australia and New Zealand. Some of the children, many of them now young adults, have been revisited in 2007 to further record their progress.

Rob Last
Vision Australia – Early Childhood Educator
69-71 Atherton Road
Oakleigh Victoria 3166 AUSTRALIA
E-mail: roblast@iinet.net.au

Transition Strategies

Strategies which may assist success when choosing appropriate educational settings and when considering the importance of communication within that setting

Rob Last
Early Childhood Educator
Teacher of the Deaf and Vision Impairment
Vision Australia
69-71 Atherton Rd
Oakleigh Victoria 3166
AUSTRALIA
E-mail: roblast@iinet.net.au

Madelene Rich
Parent
PO Box 256
Seaford Vic 3198
AUSTRALIA
E-mail: ctos@teksupport.net.au

In choosing an educational setting that seems to meet the communication needs of your child there may be many considerations that need to be addressed.

Following are suggestions and prompts as to ideas and issues that parents may wish to consider when embarking on this process or when next meeting with the teachers in their child's current school placement or when considering a new school placement for their child.

1. Draw up a detailed plan and use your professional contacts to assist you with this. Discuss with them your child's strengths and weaknesses, which will then form the basis for discussions with the school and how they might support these needs. This information is also relevant when completing funding applications for children with additional needs.
2. Commence the planning early, at least one year prior to the school placement whether it's preschool, primary school, specialist school, a specialist unit in a regular school, secondary school, university or another tertiary education setting.
3. **Vision Issues:** Arrange a thorough vision assessment:
 - what can your child see?
 - what size pictures can they see at what distance?
 - what size print can they see comfortably?
 - what are the best positions for seeing in the classroom?
 - what can my child see in the outdoor environment?
 - what can my child see in the mid and long distance?
 - what computer modifications may be needed, i.e. keyboard modifications, suitable mouse and screen size?

4. **Hearing Issues:** Arrange a thorough hearing assessment:
 - have the best hearing aids been fitted?
 - has an up-to-date FM system been provided?
 - what can your child understand in a good listening environment?
 - what can your child understand in a poor listening environment?
 - is the cochlear implant providing the best auditory information needed and what listening environments are the most appropriate?
 - is the FM system provided for the cochlear implant appropriate for the school setting?
5. **Physical Issues:** Arrange a thorough Occupational Therapy assessment:
 - what are my child's fine motor skills and challenges?
 - what are my child's gross motor skills and challenges?
 - what are the best seating arrangements for my child?
6. **Mobility Issues:** Once a school has been decided upon arrange an Orientation and Mobility assessment of the school's indoors and outdoors areas
7. **Eating and Personal Care Issues:**
Does the school understand the process and management of nutritional needs and personal care issues and are they prepared to support these needs?
8. **List the possible educational placement options.**
9. **Visit each of these options** and ask someone to attend with you to act as an advocate.
10. **Question the school's understanding** of the communication options appropriate to your child and their ability to incorporate these options into the curriculum. These options may be:

Visual communication:

- | | |
|--|------------------------------------|
| - Signed English | - American Sign Language |
| - Makaton (a key word signing system) | - Board maker |
| - Compic | - Photo Books |
| - Picture books | - Object symbols |
| - Real objects | - Electronic communication devices |
| - Note taker | - Computer needs such as a laptop |
| - Whiteboards that print out the teacher's notes | |

Communication through audition:

- | | |
|----------------|---------------------------------|
| - Hearing aids | - Cochlear implant |
| - FM systems | - In-room amplification systems |

11. **Ask if an inclusion support worker / teacher's aid would be provided.**

Ensure the skills you require of the inclusion support worker / teacher's aide are outlined. For example: fluency in the visual language used by your child, training in special education, vigilance in the playground environment for safety reasons, social skills support, interpreter, ability to act as a notetaker and able to support nutritional and personal care needs.

Request that you be on the selection panel when choosing the inclusion support worker / teacher's aide.

12. If relevant, ask if consideration could be made to make American Sign Language the 'Language Other Than English' (**LOTE**) offered by the school.

13. Ask if the school would welcome visits from people who are deaf or vision impaired or who are deafblind, in order to provide positive **role models**.

14. Check if **Visiting Teachers** in hearing and vision would be provided. These support teachers are provided based on the child's degree of hearing and vision loss.

15. Check if a **Speech Pathologist, Occupational Therapist, Physiotherapist** could be provided on a needs basis.

16. Ask if appropriate **classroom modifications** may be made. These might be:

- reduction of glare through lighting and window modifications
- provision of suitable seating at tables and for computer use
- the provision of a footrest if required
- computer modifications to suit the specific needs of the child
- the provision of a tilt / slant board
- provision of contrast to maximise visual information
- the installation of an in-room amplification system
- consistent placement of classroom furniture
- provision of hooks for school bags and individual lockers

17. Question the school on their **discipline policy**, bullying policy and attitudes in regard to diversity.

18. Make a **list of the pros and cons** of each of the education options.

19. Consider the **transport options** offered by the school.

20. Strive for full **funding options**.

21. Pay close attention to your **intuition**.

22. Make a decision.

23. **Build a relationship** with the school principal, the vice principal, the classroom teacher, all staff members, the inclusion support worker / teacher's aide and the office staff.

The principal and classroom teacher should be invited to visit the preschool or school your child currently attends prior to commencement at the new placement.

24. Ensure **professional development training** for all staff members is offered in the areas of vision, hearing, physical issues and orientation and mobility. These sessions need to be ongoing as needs and issues change. Parent can also provide information on relevant seminars, conferences, workshops, newsletters, websites, reference materials, support groups and resource centres.

25. Ensure you are included in all meetings that relate to your child.

These may be:

- information meetings
- programming meetings
- goal setting meetings

26. Provide an **information sheet on your child** to be used as an information source to parents, new staff, relief teachers and teachers in training. This may be used to introduce your child and their special needs to other parents through the school newsletter.

27. **Be respectful** of the school's routines, values and culture and remember other children may have special needs that may be less obvious.

28. Remember the communication options, modifications to the indoors and outdoors and the information sessions will benefit all the teachers, children and young people in the education setting you select.

29. **Be vigilant** when changes occur in leadership and staffing.

In Conclusion

All of these suggestions may be relevant to your situation or some of these suggestions may be relevant to your situation or only one or two suggestions may be relevant to your situation. We sincerely hope that something of benefit has been gained.



PROGRAM SECTION 21

Breakout Session #4
Friday, July 27, 2007, 4:15-5:30
Balboa Bay I & II

Training for Fundraisers



Neal Stanger,
Stephen Perrault, and
Lacey Friedman

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

21 GI Training for Fundraisers *(Neal Stanger, Stephen Perrault, and Lacey Friedman)*

The CHARGE Syndrome Foundation, Inc. is heavily committed to fundraising to ensure that we can offer the services—like this conference—that are so valuable to the families and professionals we serve. This session is conducted in response to requests from our members who wish to raise funds for the Foundation. Information will be provided about the Foundation's fundraising efforts and how members can participate.



PROGRAM SECTION 22

Breakout Session #5
Saturday, July 28, 2007, 10:30-11:45
Pacific Ballroom II

Vestibular Function,
Balance, and Motor
Development



Presented by
Lori Travis and
James Thelin

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Vestibular Function, Balance, and Motor Development in CHARGE Syndrome

Lori S. Travis, Au.D., CCC-A
Clinical Audiologist

University Physicians Audiology Clinic
University of Missouri Health Care
Columbia, MO

July 27, 2007

Vestibular Function and Balance Abilities

- Many do not learn to walk independently
- Could be due to:
 - Vestibular anomalies
 - Visual deficits
 - Musculoskeletal abnormalities
 - Delayed development
- Few have had formal vestibular evaluations

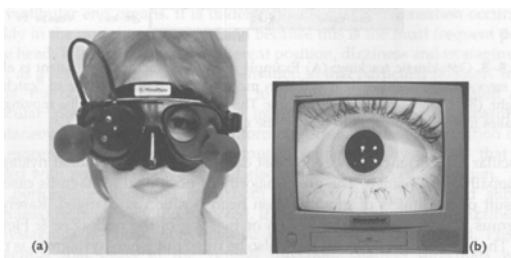
Currently Available Vestibular Assessment Methods

- Electronystagmography (ENG) or Videonystagmography (VNG)
- Posturography
- Vestibular evoked myogenic potentials (VEMP)

ENG and VNG

- Most common vestibular tests
- ENG uses electrodes placed around the eyes to measure their movement electrically
- VNG uses cameras within goggles to focus on the pupil of the eye to measure their movement

VNG Goggles



Desmond, Alan. (2004). Vestibular Function: Evaluation and Treatment. New York: Thieme.

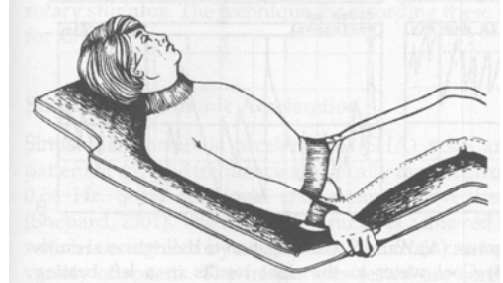
ENG and VNG

- Subtests:
 - Eye movement tasks
 - Patient follows a target with eyes
 - Positional/Positioning tasks
 - Body and head positions can elicit abnormal eye movements
 - Calorics
 - Heating or cooling ear canal to cause dizziness

ENG and VNG

- Disadvantages for testing those with CHARGE:
 - Eye movement tasks:
 - Colobomas make it difficult to follow targets
 - Positioning/Positional tasks:
 - Difficult positions; Pt must cooperate
 - Must be willing to close eyes for ENG
 - Calorics:
 - Must cooperate for 20 minutes while stimuli are being used to induce dizziness

Position for Caloric Testing



Desmond, Alan. (2004). Vestibular Function: Evaluation and Treatment. New York: Thieme.

Caloric Testing

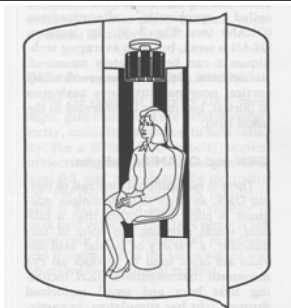


Stockwell, Charles. (2004). Introduction to ENG. Caro: GN Otometrics.

Rotary Chair testing

- ENG electrodes or VNG goggles
- Person is physically strapped in a certain position in a chair in the dark
- Chair rotates at various speeds
- Child may be able to sit in a parent's lap during the test

Rotary Chair



Jacobson, G., Newman, C., & Kartush, J. (1997). Handbook of Balance Function Testing. New York: Delmar.

Vestibular Evaluations

- Often not performed on children because:
 - Unable to tolerate the darkness
 - Unable to find or focus on target
 - Unable to obtain a proper position for the test

Vestibular Anomalies

- Vestibular anomalies (present in 80-90% of the population):
 - Deformities of the semicircular canals (SCC)
 - Partial or complete absence of all three canals in one or both ears
 - Absence of the vestibulo-ocular reflex (VOR)
- Auditory anomalies:
 - Obliteration of the oval and round windows of the cochlea

Vestibular Assessments

- Anatomy: CTs & MRIs are common
- Function: ENG/VNG, almost never done
- Effect on development gets little attention
- If SCCs absent, some function in other areas of vestibular system has been seen
- Abnormal vestibular structures ≠ Effects on balance

Vestibular Assessments

- Few studies show a relationship between vestibular problems and motor development
- Rotary chair testing was the most common test used
- Serious equilibrium difficulties and many falls during rapid head movements

Effects of Vision Abnormalities

- Abnormal vision in CHARGE: 80-90%
- Study conducted with blind and seeing infants to examine the impact of vision on early motor development
 - Those with vision had control early on
 - Those with blindness did not have control until 1 year

Communication Development

- Communication development is related to a child's ability to walk independently
- A child's ability to walk is related to the sense of balance and vestibular functioning

Thelin & Fussner (2005)

- Ability to walk independently is significantly related to the development of symbolic language
 - 94% of those with symbolic language could walk independently
 - Only 18% of those without symbolic language were able to walk independently
- Treatments could help with development of symbolic language

Research Purpose

- Gain information about:
 - How children with CHARGE have been evaluated
 - How development has progressed -
- especially when it is known that the vestibular mechanism is involved

Research Design

- Telephone interviews to obtain information on the child's:
 - development
 - ability to ambulate
 - ability to communicate
- Obtain parent's assessment of:
 - balance and mobility, and
 - ages at which specific proficiencies were acquired

Parent Interviews

- Parents were asked about:
 - Ability to walk and balance
 - Gross motor developmental milestones
 - Formal vestibular evaluations
 - MRI/CT scan results
 - Communication abilities
 - Hearing
 - Vision
 - Other physical or medical problems
 - Current therapies

Subjects

- Data was obtained from 9 children
- Aged 4.5 to 21 years
- 5 males, 4 females

Shift in Focus

Difficult to test population

New Information



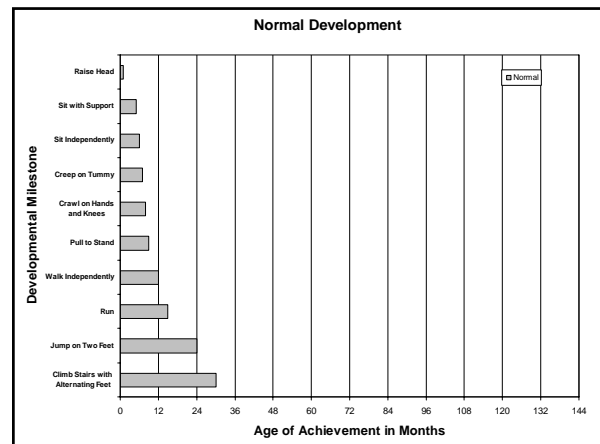
How does development, specifically walking independently, relate to communication abilities??

Comparison of Data

- Development norms vs. Development in children with CHARGE
- What is the relationship between:
 - Gross motor development and abilities and
 - Communication development

Ten Milestones Used to Assess Gross Motor Development

- Raise head
- Sit with support
- Sit independently
- Creep on tummy
- Crawl on hands and knees
- Pull to stand
- Walk independently
- Run
- Jump on two feet
- Climb stairs with alternating feet



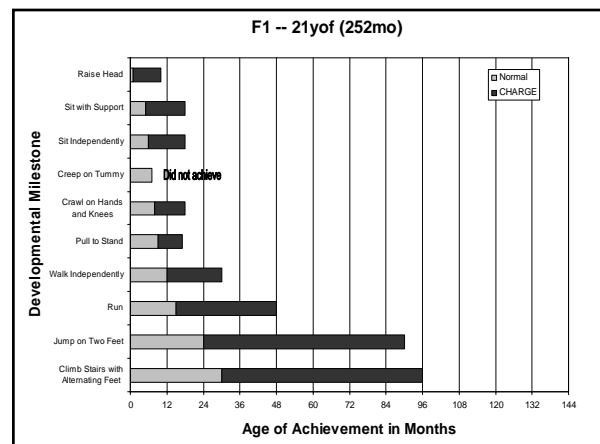
Proficient Language Group

F1 – 21 year-old female

- Diagnosed at age 14 years
- Physical abilities:
 - Walks well
 - Runs
 - Cannot ride a bike
 - Plays hockey and basketball
 - Poor balance
- Graduated high school at 18
- Communication:
 - Speaks

F1 – 21 year-old female

- Hearing:
 - Profound loss in one ear, moderately-severe to profound in other
 - Began wearing hearing aids around the age of 2 years
 - Wears her hearing aid all day and does very well
- Vision:
 - Blind in one eye, impaired central vision, myopic, and astigmatism in other eye
 - Began wearing glasses around the age of 10-12 years
 - Wears glasses and does well

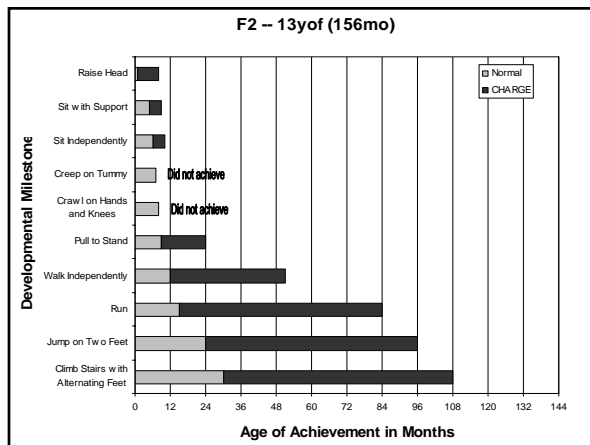


F2 – 13 year-old female

- Diagnosed at 5 days old
- Physical abilities:
 - Walks fairly well – flat footed and wide based
 - Runs and skips
 - Motion from sitting to standing is difficult
 - Has trouble with 1 footed jumps or standing for more than 3 seconds
 - Balance has become good within the last 2 years
- Is in 5th grade at a residential deaf school; repeated K and 1st grades
- Communication:
 - Proficient sign language

F2 – 13 year-old female

- Hearing:
 - Profound loss in both ears
 - Wore aids from 1997-2000, but not very often
 - Stopped receiving benefit from aids after a while, so no longer wears them
- Vision:
 - Retinal coloboma in one eye, not impacting major visual field
 - Mother wonders if she has cortical visual impairment (CVI)
 - Wears glasses for slight farsightedness (runs in family)



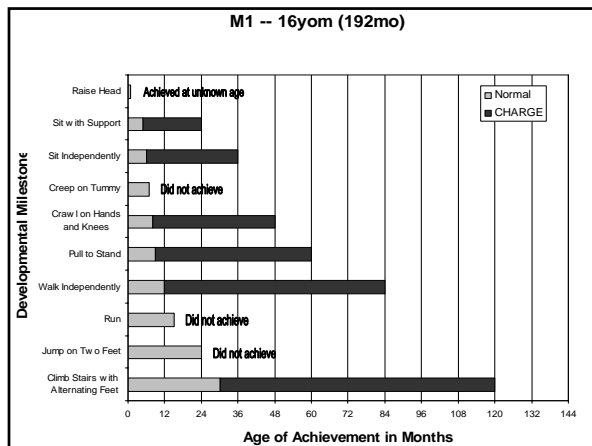
Minimal to No Language Group

M1 – 16 year-old male

- Diagnosed at 5 weeks old
- Physical abilities:
 - "Toddler walk"
 - Can walk without support on level ground
 - Holds onto someone so as not to trip on unsteady ground
- Attends school
- Communication:
 - No spoken language
 - Uses Picture Exchange Communication System (PECS) for familiar wants and needs
 - Leads
 - Uses approximately 3 signs, understands 20-25 signs

M1 – 16 year-old male

- Hearing:
 - Severe to profound loss in both ears
 - Has had a cochlear implant for 3 years for one ear, does not wear an aid on the other ear
 - Does not understand well
- Vision:
 - Colobomas cover the macula and fovea in one eye, and skirts the macula in the other eye
 - Has worn glasses since the age of 6 months
 - Has poor vision in one eye, has "baseball cap vision" and is very nearsighted in the other eye

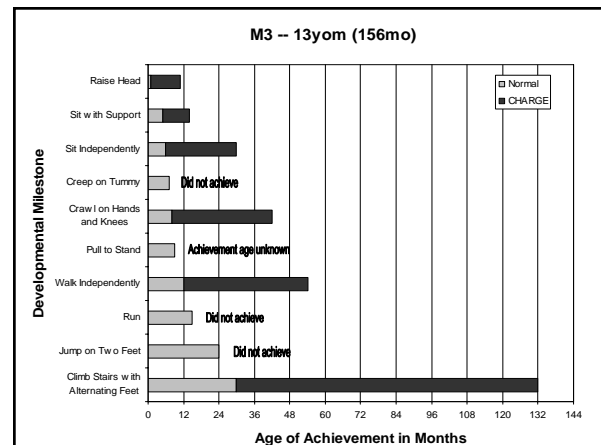


M3 – 13 year-old male

- Diagnosed at 2.5 months of age
- Physical abilities:
 - Does not move quickly but does walk independently on even surfaces
 - Has foot braces and scoliosis
 - Does not run
 - Uses a rehabilitation stroller for walking longer distances
 - Can now stand well in one position without losing balance
 - Loses his balance easily but catches himself well, rarely falls
- In 5th grade, mostly in special education setting

M3 – 13 year-old male

- Communication:
 - Makes some speech-like sounds
 - Uses BigMacs – understands the cause and effect
- Hearing:
 - No known hearing loss
 - Possibly some auditory processing issues, but unable to test for this
 - Uses an FM system at school which helps with attention
- Vision:
 - Has bilateral colobomas located centrally all the way to the macula and nerve
 - Legally blind but is functionally ok, compensates well
 - Can orient and move around

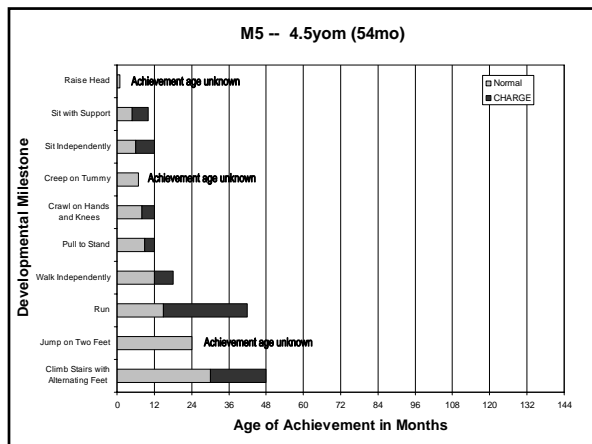


M5 – 4.5 year-old male

- Diagnosed at 1 week old
- Physical abilities:
 - Walks with wide gait and sways side to side
 - Runs
 - Does not do well on uneven surfaces
 - Loses balance easily
 - Can catch self but falls as well
- Attends a center-based preschool program part-time with other severely multiply involved children

M5 – 4.5 year-old male

- Communication:
 - Leads
 - Tried PECS
 - Tried sign – uses few signs minimally and not consistently
 - Understands more than he produces
 - Recently began responding to his name by coming when called
- Hearing:
 - Profound loss in one ear, moderate in other
 - Generally refuses to wear hearing aids
 - Will go through phases of wearing them at school or in therapy, but is a struggle to get them in
- Vision:
 - Complete colobomas in both eyes in bottom of visual field, nearsighted
 - Functionally he does well as he looks out the top of his eye
 - Has glasses but refuses to wear them



Language Comparisons

- Proficient Language (sign or spoken)
 - 5 of 9 subjects
 - Walked sooner as a group
 - Developmental spurts (exception of M5 for whom this is also true)
 - "Walk independently" milestone was dividing line of development

Language Comparisons

- Minimal to No Language (leads, BIGmack, PECS, sign)
 - 4 of 9 subjects
 - 3 of the 4 were more delayed in their *earliest milestones* than the proficient language group
 - Most severely involved kids
 - ***Slow but steady progress*** across all milestones
 - More "did not achieve" than proficient group
 - 1 child was an exception; see next slide

Language Comparisons

- The child was an exception for 2 reasons:
 - Developed fastest and closest to normal compared to the others
 - But, has very minimal language (very few signs, leads)
- What factors might explain this?
 - Vision
 - Hearing loss
 - Autistic-like qualities

Summary

- 2 main groups of children
 - More typical gross motor development: walk and have language
 - Significantly delayed gross motor development: walk and have minimal to no language
- Persistent question in the CHARGE community:
 - Are they delayed or does their development stop?
 - Appears as if they are mostly delayed
 - It may take them a while to get it, but they tend to eventually get it

The Vestibulo-Ocular Reflex and CHARGE SYNDROME

James W. Thelin

o0o

Department of Audiology & Speech Pathology
University of Tennessee

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 28, 2007

Collaborators in Research on CHARGE Syndrome at the University of Tennessee

Julie Stephens Jackson
Katie A. Vaden
Donna Fisher Smiley
Jill Fussner Maddox
Emily C. Barker
Holly A. Beason

Kristina Keller Walton
Lori A. Swanson
Lori Ruhl Travis
Nancy Steele
Emily King
Sarah Krivenki

Communication & Physical Development

From the literature:

- *"Communication develops as part of an interdependent system with physical development, thus development in one area facilitates the development of the other."*

(Bates, 1979)

- We asked "With the large number of physical and sensory problems present in CHARGE, how is communication affected? Are there disorders that have more of an effect on communication?"

Which of the features in CHARGE (major, minor, and other features) might contribute to the *INABILITY to walk independently?*

In the three following slides, features that are likely to contribute are shown in *RED* and those that could possibly contribute are shown in *BLUE*.

CHARGE Diagnostic Criteria (Revised 1998)

Major Features

- Coloboma: 80-90%
- Choanal atresia: 50-60%
- Cranial nerve dysfunction
 - N I (smell): frequent N VIII (hearing & balance): 75-80%
 - N VII (face): 40%+ N IX/X (swallowing): 70-90%
- Characteristic CHARGE external ear: 90%
- Middle ear anomalies: 90% ?

CHARGE Diagnostic Criteria (continued)

Minor Diagnostic Criteria

- CHARGE face: >50%
- CHARGE hand: 50%
- Congenital heart: 75-85%
- Cleft lip/palate: 20-30%
- Hypotonia: frequent
- Renal anomalies: 40%
- TEF/esophageal atresia: 15-20%
- Middle ear infections/PE tubes: >80%
- Short stature: common
- Growth hormone deficiency: rare

CHARGE Diagnostic Criteria (continued)

Other Common Findings

- | | |
|----------------------------------|--------------------------|
| -Microcephaly | -Webbed neck |
| -Agenesis of the corpus callosum | -Abdominal wall defects |
| -Apnea | -Scoliosis |
| -Seizures | -Limb/skeletal anomalies |
| -Laryngomalacia | -Autistic-like behavior |
| -Thymic/parathyroid hypoplasia | -Behavior problems |
| | -Nipple anomalies |

Communication & Physical Development

(Thelin & Fussner, 2005)

- Prevalence of significant physical involvement for all participants:

- Vision loss	100%
- Hearing loss	94%
- Swallowing/feeding problems	87%
- Hypotonia	81%
- Choanal atresia	74%
- Heart problems	74%
- Gastrostomy tube	58%
- Facial problems	55%
- Growth deficiency	55%
- There was no significant relation between the acquisition of symbolic language and any of these disorders

Factors Related to the Acquisition of Symbolic Language (Thelin & Fussner, 2005)

- (1) Success in the treatment of hearing loss with amplification
- (2) The ability to walk independently (possibly as a result of many factors including vision and vestibular function) and
- (3) Communication training initiated before 3 years of age.

Communication & Mobility

From the literature:

- *Approximately one-half of youth who are deaf-blind do not use symbolic communication.*
(Collins, 1993)
- *Approximately one-half of youth who are deaf-blind **AND** do not have symbolic communication **ALSO** do not walk independently.*
(Petroff, 2001)

How could the "ability to walk independently" affect the acquisition of symbolic language?

1. The inability to walk independently may be due to significant neurologic involvement or growth deficiency.
2. The inability to walk may prevent to individual from positioning himself/herself within a "COMMUNICATION BUBBLE" that enables optimal visual and auditory communication.

Communication & the Communication Environment

From the literature:

Communication Bubbles:

Regions around an individual in which a talker or signer can be heard and seen optimally or acceptably.

-- The bubbles may be different for hearing and vision and will change with background noise and lighting.

-- For individuals with deaf-blindness, communication may be greatly compromised if the talker or signer is not within the bubbles.

(Davenport, 2002)

BALANCE & Mobility in CHARGE

- The ability to walk independently is significantly related to the ability to acquire symbolic language.
- Children with CHARGE have vestibular anomalies and visual deficits that may distort visual images when the head is stationary and when the head is in motion.
- These disorders may contribute significantly to the inability to walk independently.

Balance & Mobility

Balance and mobility depend on:

- Proprioception,
- Vision, and
- Vestibular Function (*assessed with ENG, VNG, rotary chair tests, head movement tests*)

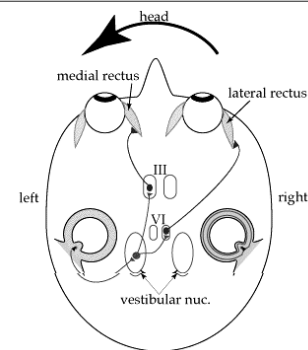
Adequate function of 2 of these 3 systems is required to maintain balance. Better function is required for mobility.

Vestibulo-Ocular Reflex (VOR)

- Purpose: *To stabilize visual images while the head is in motion; "steady cam".*
- Structures: *Linkage of the semicircular canals that sense angular acceleration, neurons in the brainstem, eye muscles, and eyes that track visual images; coordinates head and eye movement.*
- Measurement of the VOR: *Electrodes next to the eyes or cameras are used to record eye movements when the head is moved; ENG and VNG.*

Vestibulo-Ocular Reflex Arc

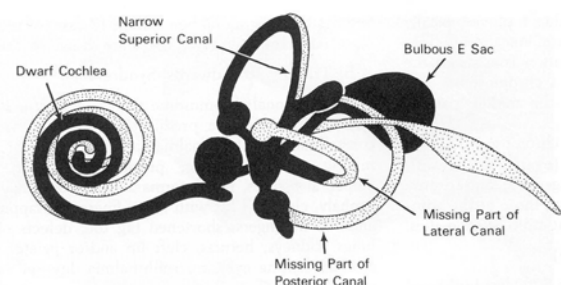
Kaiser & Potter, 2006



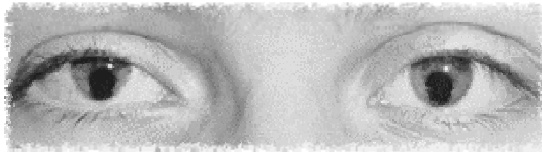
VOR Abnormalities in CHARGE

- Semicircular canals may be absent or malformed.
Motion of the head may be sensed inappropriately or not at all.
- Colobomas of the iris, retina, and/or optic nerve are common.
Visual images may not be seen :
 - *because they are in a void or*
 - *because they may disappear and reappear at unexpected locations when the head or image is in motion.*

Cochlear and Vestibular Labyrinths: Mondini's Dysplasia

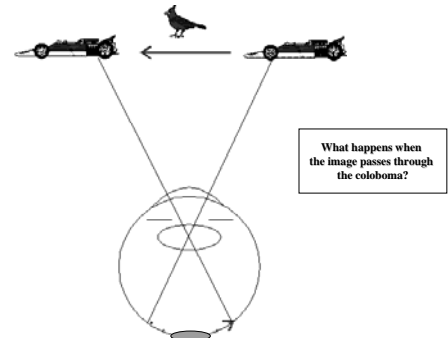


Coloboma of the Iris



Motion Detection with a Fixed Eye

Kaiser & Potter, 2006



Motion Detection with the Eye in Motion

Kaiser & Potter, 2006

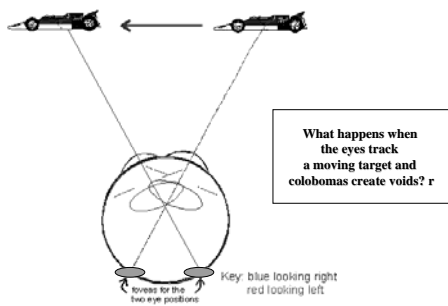
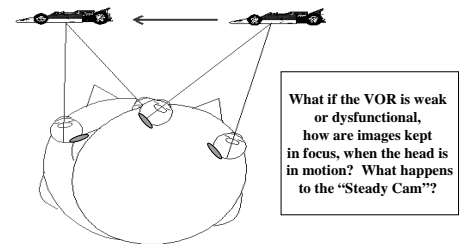


Image Stabilization & Detection of Motion with the Head in Motion

Kaiser & Potter, 2006



Eye Dynamics/Intellinetx Videonystagmography (VNG) Infrared Goggles



UT VNG VOR Test

- VNG goggles are placed over the eyes and held in place by the examiner.
- With the eyes in total darkness, the head is moved into different positions to determine if nystagmus can be provoked.
- If nystagmus is provoked, it means that the semicircular canals have directed the eyes to move and that the VOR is at least partially present.

UT VNG VOR Test

- Test takes ~3 minutes and requires minimal cooperation
- Has the promise to determine if the VOR that is responsible for stabilizing visual images while the head in motion.
- The test might be used on the very young who are incapable of the cooperation required for clinical ENG/VNG tests of vestibular function.

INFORMATION

- James W. Thelin, Ph.D.
Department of Audiology & Speech Pathology
University of Tennessee
578 South Stadium Hall
Knoxville, TN 37996-0740
Tel: 865-974-1796 Email: jthelin@utk.edu
- CHARGE Syndrome Foundation, Inc.
www.chargesyndrome.org



PROGRAM SECTION 23

Breakout Session #5
Saturday, July 28, 2007, 10:35-11:45
Pacific Ballroom III

STRATEGIC CONFLICT
MANAGEMENT PROCESES IN
SPECIAL EDUCATION
SETTINGS:
IEP Facilitation, Resolution
Meetings, and Mediation



Presented by
John Reiman

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Strategic Conflict Management Processes in Special Education Settings

Costa Mesa, CA
John W. Reiman, Ph.D.
Associate Director, National
Consortium on Deaf-Blindness
Mediation Specialist, CADRE*
July 27-July 29, 2007

*CADRE is the OSEP-funded
National Center on Dispute
Resolution in Special Education.

The materials presented herein are adapted
from presentations made nationwide by
CADRE.

2

"Seek first to
understand, then to be
understood."

Stephen Covey, "Habit 5"
Seven Habits of Highly Effective People

3

"Listening is probably the most
cost effective element of a
conflict management system."

Mary Rowe

4

Topics for Today

- ▣ Positions and Interests (brief intro)
- ▣ Dispute Resolution Systems in
Special Education & The CADRE
Continuum of Processes and
Practices
- ▣ Process Comparisons

5

Assumptions...

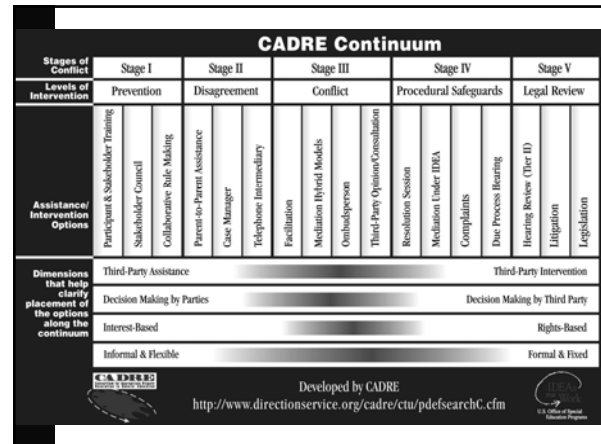
- ▣ Different cultures have differing
perspectives on conflict management
- ▣ Conflict is a healthy reflection of a
diverse and changing society
- ▣ Most parent/school relationships are or
can be positive and mutually respectful
- ▣ Skills can be acquired for productive
relationships (personnel prep)

6

Assumptions...

- ❑ Early opportunities exist to collaboratively address differing viewpoints
- ❑ Adversarial processes should be reserved until other options have been exhausted
- ❑ Adversarial processes can have high personal, relational and financial costs

7



What is IEP Facilitation?

- ❑ All IEP meetings benefit from skilled and capable facilitators who can assist the team in crafting agreements
- ❑ A facilitator helps keep members of the IEP team focused while addressing conflicts and disagreements that may arise during the meeting
- ❑ A facilitator will use communication skills that create an environment in which the IEP team members can listen to each member's point of view

9

Who are the Facilitators?

- ❑ Team member
- ❑ Parent, trained parent advocate, or support person
- ❑ Students (when appropriate)
- ❑ An independent, external, trained facilitator not affiliated with the team or school district

10

Role of the Facilitator

- ❑ Helps create an overall agenda and assists in generating ground rules
- ❑ Guides the discussion by keeping the team's energy centered and helps to identify underlying interests
- ❑ Assists the team to resolve conflicts and disagreements that arise
- ❑ Helps to maintain open communication

11

Role of the Facilitator

- ❑ Helps team members develop and ask clarifying questions
- ❑ Helps to keep team members on task and within the time allotted
- ❑ Maintains impartiality and does not take sides, place blame or render judgments
- ❑ Does not impose a decision on the group

12

Potential Benefits of a Facilitated IEP Meeting

- ❑ Supports full participation of all parties
- ❑ Builds and improves relationships
- ❑ Insures meeting remains student-focused
- ❑ Models effective communication and listening
- ❑ Clarifies points of agreement and disagreement

13

Potential Benefits of a Facilitated IEP Meeting

- ❑ Provides opportunities to address conflicts
- ❑ Encourages identification of new options
- ❑ Costs less than more formal proceedings
- ❑ Typically less stressful than formal proceedings
- ❑ Supports better follow through and follow-up
- ❑ Serves as the IEP meeting

14

What is a Resolution Meeting?

- ❑ A Resolution Meeting is a new conflict resolution procedure established under the Individuals with Disabilities Education Improvement Act of 2004 (IDEA 2004).
- ❑ A Resolution Meeting occurs within 15 calendar days of the filing of a due process complaint, unless the parent and district agree in writing to waive the meeting or agree to mediation.

15

What is a Resolution Meeting?

- ❑ A Resolution Meeting affords parents and the school a chance to work together to resolve a disagreement and avoid a due process hearing.
- ❑ A Resolution Meeting includes the following participants: the parents, an LEA (local educational agency) representative who can make decisions, and any Individualized Education Program (IEP) team member who has relevant information about the issue that is being discussed. School districts may not bring an attorney to the Resolution Meeting unless the parents bring an attorney.

16

Benefits of Participating in a Resolution Meeting

- ❑ An important "last chance" to talk things over
- ❑ Less adversarial than a due process hearing
- ❑ Opportunity to develop a mutually agreeable solution
- ❑ Possibility of repaired communication and relationship
- ❑ Parents and school decide which members of the IEP team will attend the meeting
- ❑ May result in legally binding agreement enforceable in court

17

Cautions about the Resolution Meeting

- ❑ Going in unprepared reduces the chance of a durable agreement
- ❑ Some situations could benefit from a third party facilitator
- ❑ No guarantee that an agreement will be reached
- ❑ Can be emotional
- ❑ Questions exist related to confidentiality of process

18

Mediation Under IDEA '04

Conducted by a mediator who is:

- ❑ Qualified
- ❑ Trained in effective mediation techniques
- ❑ Knowledgeable in laws and regulations
- ❑ Impartial
- ❑ Not employee of LEA, SEA or state agency that is providing direct service to the child
- ❑ No personal or professional conflict of interest
- ❑ Random selection or both parties agree

19

Mediation Agreement 615(e)(1), (2)(F)

- ❑ Mediation is available to resolve any matter, even before requesting a due process hearing (DPH).
- ❑ Mediation agreement must be in writing and signed by the parent and agency representative.
- ❑ Agreement is legally binding and enforceable by any state court.
- ❑ Mediation process is confidential and may NOT be used as evidence in subsequent legal action.

20

The Mediation Process

- ❑ Setting the Stage
 - ❑ Prepare environment
 - ❑ Explain expectations & process
 - ❑ Agreement to Mediate
 - ❑ Confidentiality
 - ❑ Role of The Mediator
- ❑ Uninterrupted Time
 - ❑ Invite each person to talk without interruptions
 - ❑ Participants describe situation and their perception of the issues (initial expression and release of emotions around situation)

21

The Mediation Process

- ❑ Identifying the Issues
 - ❑ Identify, clarify, summarize main issues
 - ❑ Get agreement on issues
- ❑ Discussing the Issues
 - ❑ Listen for common ground, negotiable issues, points of disagreement
 - ❑ Encourage participants to talk to each other

22

The Mediation Process

- ❑ Generation of Potential Solutions
 - ❑ Assist in brainstorming possible options for each discrete issue
 - ❑ Evaluate potential options
- ❑ Agreement Building
 - ❑ Narrow list of viable options
 - ❑ "Reality Check"
 - ❑ Assist in negotiation process
 - ❑ Record agreement if appropriate on some or all issues

23

Cultural Competence and Diversity

The Platinum Rule:

"Do unto others as they would have you do unto them."

Tony Alessandra

24

Cultural Competence and Diversity

- ❑ Some individuals do not welcome involvement of government
- ❑ Some cultures fear challenging professionals
- ❑ Families might not understand mainstream western beliefs about "parent-educator partnerships"
- ❑ Actions and words don't always have the impact we intend

25

Cultural Competence and Diversity

- ❑ Determine most appropriate method of communication (e.g., in-person, email, phone)
- ❑ Be aware of personal biases and assumptions based on how a person dresses, speaks, acts, etc.
- ❑ Insist that mediators have no perceived and/or real conflicts of interest

26

Cultural Competence and Diversity

- ❑ Recognize that many people do not communicate in a linear fashion nor "stick to the subject at hand"
- ❑ Avoid language or assumptions that perpetuate stereotypes
- ❑ Allow time for reflection, don't always fill silent spaces
- ❑ Engage community leaders and cultural liaisons in outreach and model definition

27

Cultural Competence and Diversity

- ❑ Arrange the room and seat the participants in a respectful and functional manner
- ❑ Permit joint and individual meetings as appropriate for saving face, venting, consultation with advisors

28

Student Participation

The student should be present and participate as much as possible in any meeting regarding their services.

"Nothing about me
without me."

29

Advantages Related to Student Participation:

- ❑ Vital opportunity for student to be understood
- ❑ Student's preferences taken directly into account
- ❑ Student learns to articulate needs to service providers
- ❑ Student empowerment
- ❑ Elevates civility of process
- ❑ Encourages positive, strength-based discussions
- ❑ Creates new relationships and linkages
- ❑ Ensures presence of key person in design and implementation of effective agreement

30

Challenges Associated with Student Participation:

- ❑ Competency to participate
- ❑ Disagreement between parents and student
- ❑ Coercion and forced agreements
- ❑ May limit topics of discussion
- ❑ Potential for student to accept unreasonable levels of responsibility

31

Key Components of Any Written Agreement

- ❑ Use clear, simple and positive language.
- ❑ Frame actions as to what participants **WILL** do, not what they **WON'T** do.
- ❑ Be specific as to names, places, dates, times, amounts, and actions.
- ❑ Use language that is future oriented. Avoid reference to past problems or blame.

32

Key Components of the Agreement

- ❑ Avoid contingencies (e.g., "The school will do **xyz** **if** the parents do **abc**"). Each discrete activity should stand on its own.
- ❑ Use impartial, non-judgmental language. Write "Richard agrees to..." rather than "Richard should..." or "Richard must..."
- ❑ Provide for the future. Encourage participants to develop a process for monitoring the agreement and creating modifications as needed.

33

Contact Info

John W. Reiman, Ph.D.

Associate Director, National Consortium on Deaf-Blindness

Mediation Specialist, CADRE*

reimanj@wou.edu

503-838-8776

503-838-8821 tty

504-838-8150 (fax)

34



PROGRAM SECTION 24

Breakout Session #5
Saturday, July 28, 2007, 10:35-11:45
Pacific Ballroom IV

The Behavioral Phenotype
in CHARGE



Presented by
Tim Hartshorne

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

The “B” in CHARGE

Tim Hartshorne
Central Michigan University
tim.hartshorne@cmich.edu

Costa Mesa, CA
July 28, 2007

A bit of History

- 1979 – Hall identifies an association
- 1981 – The association is called CHARGE
- 1993 – First US conference – nothing on behavior
- 1995 – Bernstein presented on behavior in adolescents at 2nd US conference
- 1996 – Workshop at 2nd Australasian conference on behavior by Williams, Hartshorne, and Hartshorne
- 1997 – Bernstein presents data at 3rd US Conference

More History

- 1998 & 1999 – Hartshorne presents on “behavior as communication” at Australasian and US conferences
- 1999 – Fernell, et al., Swedish study on autism in CHARGE
- 2003 – Behavior Symposium at 6th US Conference – nine papers presented
- 2004 – Hartshorne & Cypher paper on behaviors
- 2005 – American Journal of Medical Genetics

Personal History

- 1989 – Jacob is born with CHARGE
- Odd, autistic-like behaviors in first two years. Introduced to the impact of deafblindness.
- Looked at parent-professional relationships and impact on parents
- As behavior problems became evident, figured out that a psychologist was needed

Toward an Understanding of Behavior in CHARGE Syndrome

- Understanding the syndrome
- Considerable observation of many children
- Research to identify commonalities
- Research to compare with other syndromes
- Consensus building

Understanding the Syndrome

Most genetic syndromes have behavioral features

- What we have learned from parents
- Early best guesses
 - Deafblindness
 - Vestibular dysfunction
 - Communication difficulties/frustrations
 - Early experiences (pain, attachment)
- Critique and Challenge

Observations of Children

- Conferences
- Parent descriptions and questions
- Consultations
- Video

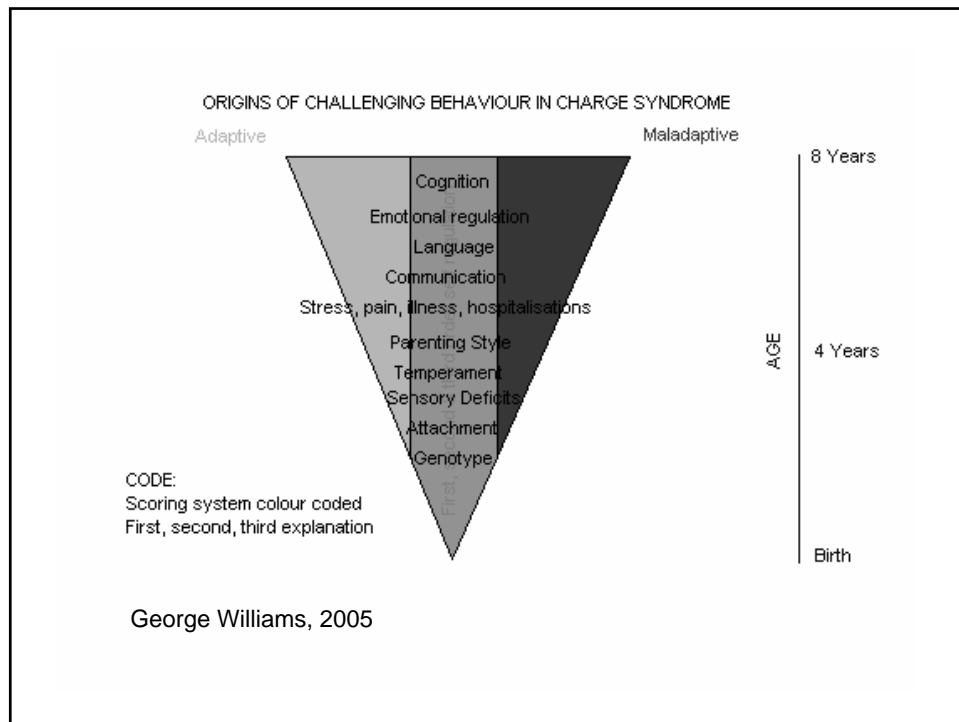
Research

“extreme preference for certain toys, people, food, etc.”

“restricted range of interests and/or pre-occupation with one narrow interest.”

“significant difficulty in ability to make same age friendships.”

Hartshorne, T. S., & Cypher, A. D. (2004). Challenging behavior in CHARGE syndrome. *Mental Health Aspects of Developmental Disabilities*, 7(2), 41-52.



Attachment Problems

- May arise from low or absent levels of:
 - “visual, vocal or other forms of reciprocity occurring from the neonatal days on.”
- A huge issue for CHARGE babies from the first hours onwards with sensory deficits, medical issues, pain, separation, parent shock, etc.
- 20% of parents had delayed bonding, and nearly a half reported delayed attachment from the child.

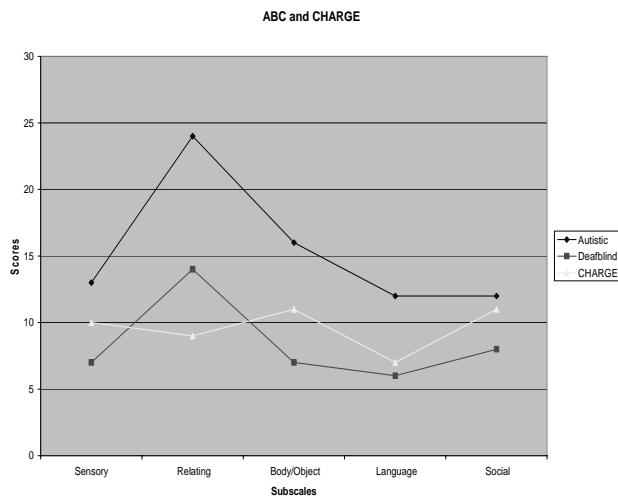
Reda, N. M. & Hartshorne, T. S. (in press). Attachment, bonding, and parental stress in CHARGE syndrome. *Mental Health Aspects of Developmental Disabilities*.

Sensory Deficits

- Hearing – sensori-neural hearing loss
- Vision – coloboma
- Smell – anosmia
- Taste – prefer strong tastes
- Tactile – defensiveness
- Vestibular – missing semi-circular canals
- Proprioceptive – muscle weakness

Typical Deafblind Behavior

- | | |
|---------------------------|--------------------------|
| • Eye pressing | • Vocal tics |
| • Finger flicking | • Feces smearing |
| • Rocking | • Lining things up |
| • Tapping body/objects | • Extreme preferences |
| • Self-injurious behavior | • Darting/running off |
| • Mouthing objects | • Learned helplessness |
| • Tactile defensiveness | • Submissive |
| • Clinging | • Stare at lights |
| • Spinning | • Inappropriate vocalize |



Hartshorne, T. S., Grialou, T. L., & Parker, K. R. (2005). Autistic-Like Behavior in CHARGE Syndrome. *American Journal of Medical Genetics*, 133A, 257-261.

How do you parent a child with CHARGE?

- Do you use time out?
- Do you use food as a reinforcer?
- Can you explain the reason for behavior?
- Can you say “no”?
- Can you manage your level of stress?

Parenting Style

- *The Malaise Inventory*. This 24-item questionnaire was developed by Rutter, Tizard, and Whitmore (1970) as a brief measure of mother's mental well-being. The items refer to emotions and somatic complaints. In a sample of 87 parents of children with CHARGE, one third achieved a clinically significant score.

Hartshorne, Dailor, Williams, Papadopoulos, Doyle & Brandt (2005). Sleep disturbance in CHARGE syndrome. Society for the Study of Behavioural Phenotypes, Cairns, Australia.

Parenting Style

The *Parenting Stress Index Short Form* (PSI-SF) (PSI, Abidin & Ona, 1995) is a 36-item self-report measure that was used to identify stressors that are being experienced by the family that relate to parenting. Twelve of 25 parents of children with CHARGE achieved a clinically significant score.

Reda, N. M. & Hartshorne, T. S. (in press). Attachment, bonding, and parental stress in CHARGE syndrome. *Mental Health Aspects of Developmental Disabilities*.

Pain, Illness, Hospitalizations

- Delays in coming home from birth (with mother to more than 13 weeks)
- Number of surgeries (1/4 had 13 or more)
- Number of hospitalizations (1/4 had 13 or more)
- Sleep disorder (58% a clinical disorder)
- Stress?

SLEEP

Scale	Mean	SD	% clinical
• Initiating/Maintaining	62.05	15.31	
• Breathing	59.63	15.21	
• Arousal	48.57	5.87	
• Transition	57.08		13.77
• Somnolence	51.76	11.62	
• Hyperhydrosis	49.91	10.01	
• Total	59.29	13.11	57.5%

Mother well being was associated with the child's sleep.

Hartshorne, Dailor, Williams, Papadopoulos, Doyle & Brandt (2005).
Sleep disturbance in CHARGE syndrome. Society for the Study of
Behavioural Phenotypes, Cairns, Australia.

Stress

- Child
- Mother
- Family
- School

Communication/Language

Makes reactions or noises or behaviors which can be difficult to interpret	20	16.1%
Uses behaviors such as gestures, sounds, body movements	12	9.7%
Uses single words, signs, picture symbols, or object symbols to represent basic needs	15	12.1%
Uses some 2- to 5-word phrases and sentences using speech, signs, picture symbols, etc.	17	13.7%
Uses verbal or sign language in complete sentences	59	47.6%

All children were 4 or older

Dunn Conceptual Model

Neurological Threshold Continuum	Responds in ACCORDANCE with threshold	Responds to COUNTERACT the threshold
HIGH (habituation)	Poor Registration	Sensation Seeking
LOW (sensitization)	Sensitivity to Stimuli	Sensation Avoiding

Sensory Profile

- Four factors on Sensory Profile significant
 - Low endurance/tone
 - Poor registration
 - Fine motor/perceptual
 - **Sensation seeking**
- Suggestive of a Regulatory Disorder

Hartshorne, T. S., Grialou, T. L., Russ, J. M., Nicholas, J., & Dunn, W. (2004). Executive dysfunction and sensory processing in CHARGE syndrome. Society for the Study of Behavioural Phenotypes, Barcelona, Spain

Cognitive Ability

- *Adaptive Behavior Evaluation Scale* with 100 Children with CHARGE
- Showed a higher range of ability than once thought: 54 had scores > 70 (Mean=100; SD=15)
- Those who walked earlier, had fewer medical problems, and had better hearing and vision scored higher on the ABES.
- The majority of the variance in ABES scores was explained by age at walking.

Salem-Hartshorne N, Jacob S. 2004. Characteristics and development of children with CHARGE association/syndrome. *Journal of Early Intervention* Vol 26(4): 292-301.

EXECUTIVE FUNCTION

Inhibit	64.02	(14.66)	88	46.6
Shift	65.42	(14.00)	93	57
Emotional Control	58.44	(13.06)	94	31.9
Initiate	60.93	(12.64)	87	40.2
Working Memory	62.67	(12.90)	90	12.3
Plan/Organize	60.66	(13.89)	80	41.2
Organization of Materials	52.10	(10.54)	90	12.3
Monitor	64.44	(13.40)	91	54.8
<i>Behavioral Regulation Index</i>	63.79	(13.58)	87	50.6
<i>Metacognition Index</i>	61.64	(13.00)	80	45
<i>Global Executive Composite</i>	63.00	(12.91)	79	49.4

Hartshorne, T. S., Nicholas, J., Grialou, T. L., Russ, A. M. (2007). Executive function in CHARGE syndrome. *Child Neuropsychology*, 13, 333-344.

Life-Span Developmental Approach

- Brain maturation
 - Dendritic architecture
 - Synaptic plasticity
 - Brain circuit connectivity
 - Neuronal network formation
- Environmental engagement
- Personal mastery motivation
 - Behavioral aspects
 - Emotional aspects

“A pattern of behavior that is reliably identified in groups of children with known genetic disorders and is not learned.”
(Harris, 1995)

If I behave like this, I
probably have CHARGE
syndrome

A CHARGE Behavioral Phenotype (first draft)

- Low normal cognitive functioning
- Very goal directed, persistent, and sense of humor
- Socially interested but immature
- Repetitive behaviors; increase under stress
- High levels of sensation seeking
- Under conditions of stress and sensory overload find it difficult to self-regulate and easily lose behavioral control
- Difficulty with shifting attention and moving to new activities; easily lost in own thoughts

Is it just the syndrome talking?

- Makes it seem like nothing can then be done about the behaviors. This is not true.
- On the other hand, expectations need to be realistic given the nature of the syndrome.

Diagnoses Piled On

- Autism
- ADHD
- OCD
- Tourette
- Etc.



Problems with these Diagnoses

- Create a reality that may not exist
- Lack explanatory value
- Ignore the uniqueness of the behavior
- Lead to multiple drug treatments

Factors Contributing to Challenging Behavior

- Vulnerabilities related to cognitive impairment
- Individual child's life experiences
- Factors in the immediate environment
- The specific genetic cause

Factors related to the genetic cause are relatively less under the child's control.

Generating Hypotheses From Behavioral Phenotype

- Assume enough cognitive ability to work
- It will not be easy to change the goals
- Probably does not have social support
- May have a behavior he/she wants to do
- Needs sensory input and stimulation
- Once he/she has lost it, may be hard to regroup
- A predictable routine is needed

Intervention Plan

- Understand CHARGE and the child's abilities and limitations
- High interest, appropriate, brief academic tasks
- Reinforcement system for motivation
- Functional communication training
- Sensory breaks – hang upside down
- Daily calendar system
- Recognizing the signs and responding
- Person Centered Plan and Circle of Friends

Considerations for the IEP

- PCP action plan
- What is the perfect day
- Building social supports
- Utilizing the circle of friends
- Positive behavioral supports
- What skills are required to fulfill respected roles in life and to live with dignity?



Section 25

Breakout Session #5
Saturday, July 28, 2007, 10:30-11:45
Pacific Ballroom II

Puberty and Growth
(including the response to
growth hormone)



Presented by
Jeremy Kirk

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

***PUBERTY AND
GROWTH
(including response to GH)
IN CHARGE***

Dr. Jeremy Kirk
Consultant Paediatric Endocrinologist
Diana, Princess of Wales Children's Hospital
Birmingham, United Kingdom

Puberty & Growth in CHARGE

Part of the original CHARGE diagnostic criteria:

C

H

A

R: Retarded growth and development

G: Genital abnormalities

E

Despite this there is very little published data.

PUBERTY

Birmingham Patients

- Nine patients (6 male) with CHARGE syndrome from the Endocrine Clinic at Birmingham Children's Hospital have been investigated at an average (median) age of 15.9 years (range 10.3-17.8).
- 2/7 (29%) have mutations in CHD7.

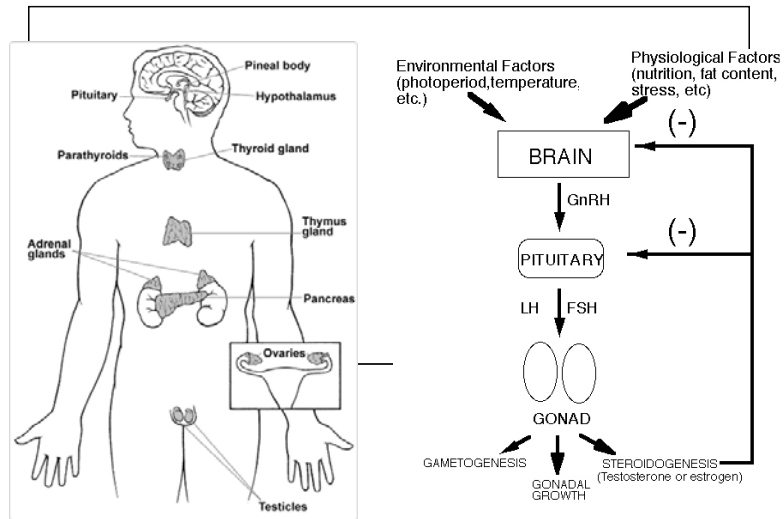
Further patients

- In addition clinical and hormonal details of a further 10 patients (2 male) with CHARGE aged 14-20 years have been obtained from other centres within the UK.

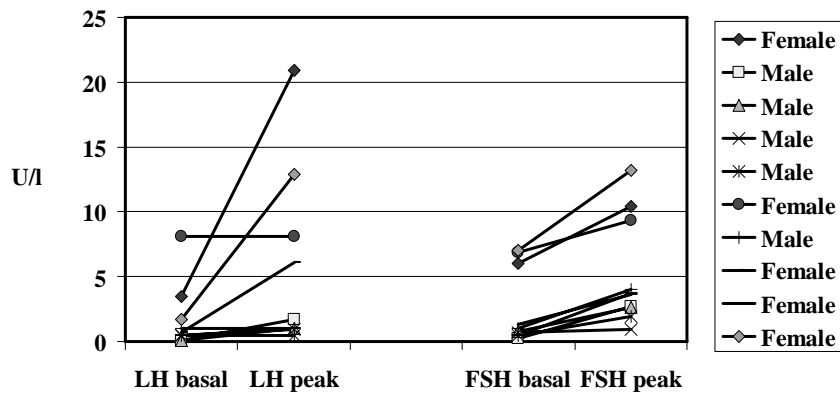
Baseline data (BCH patients)

Sex (age)	HtSDS	GH therapy	Tanner stage	Testes volume	Testosterone (pmol/l)	Oestradiol (pmol/l)	LH (IU/l)	FSH (IU/l)
F 17.8	-0.85		B2	B2		<36	3.5	6.0
M 13.4	-5.15		Y	1/1	0.6			
M 12.7	-1.18	Y	Y	1/1	<0.69		<0.1	0.2
M 13.2	0.55		Y	1/1	<0.5		0.1	0.8
M 12.8	-0.14		Y		0.9		<0.5	0.7
M 12.8	-2.99	Y	Y	3/3	<0.5		<0.5	<0.5
M 14.6	-2.28			1/1	<0.5		0.1	0.3
F 17.1	-2		B2			<73	8.1	6.9
F 10.3	-1.4	Y	B2				4.5	5.1

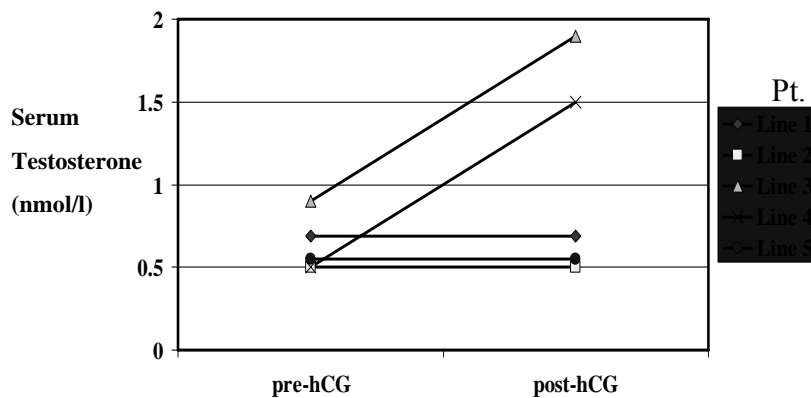
ENDOCRINE SYSTEM



LHRH testing (N=10)



hCG testing in males (N=5)



Summary of results (boys: clinical)

Boys (N=8)

- 5 had had undescended testicles (cryptorchidism)
- 5 also had evidence of micropenis (stretched penis length < 1").
- All showed failure of spontaneous pubertal development (testicular volumes < 4ml.)



Summary of results (boys: hormonal)

Boys (N=8)

- Baseline hormone levels: gonadotrophins and testosterone were low in all.
- Post hCG stimulation (N=5) testosterone rose to a mean of 1.18 nmol/l (range 0.5-1.9), with only 1 patient showing a 3-fold rise (considered normal), and 3 having little or no functioning testicular tissue.
- LHRH testing (N=5) showed pre-pubertal responses in all.

Summary of results (girls: clinical)

Girls (N=11)

- All have shown some spontaneous pubertal development (breast stage 2+), although this has usually arrested part way through.
- Only two girls have had spontaneous periods, although these have not been regular.

Summary of results (girls: hormonal)

Girls (N=11)

- Baseline hormone testing (N=5) showed detectable oestrogen levels only in three.
- LHRH testing showed (N=5) showed pubertal responses of LH in 3, and FSH in 4.
- Four girls have also had a pelvic USS performed at 17-20 years. Only 1 had a pubertal uterus, with small/non visible ovaries, with few follicles seen.

Progress

- The seven oldest patients in Birmingham have required sex hormone replacement (oral ethinyl oestradiol in the girls, and testosterone (im. depot in 3, oral in 2) in the boys.
- This has been reflected in the other patients in the UK.

Conclusions

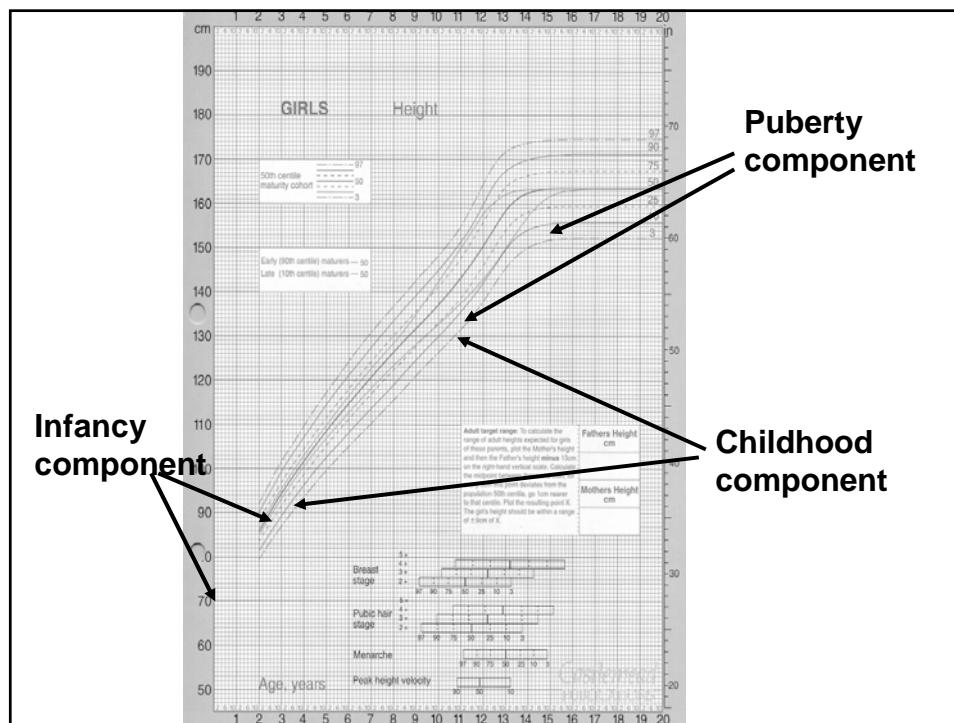
- Children with CHARGE syndrome are not necessarily short, with approximately 50% having heights within the normal range (even in those not treated with GH).
- Boys with CHARGE show clinical and biochemical evidence of gonadotrophin deficiency, and fail to enter puberty spontaneously. Although the data is limited in girls, it indicates that they show less gonadotrophin imbalance, with normal or delayed/arrested puberty.

GROWTH

NORMAL GROWTH

Normal growth occurs in 3 distinct but superimposed phases, which are under different controls:

- **INFANTILE:** first 2-3 years of life (a continuation of fetal growth). This phase is almost completely dependent on nutrition (food).
- **CHILDHOOD:** from 2 years of life until puberty. This phase is dependent on both nutrition and hormones (growth hormone predominantly, but also thyroid).
- **PUBERTAL:** from puberty onwards (starts at an average of 10 years in girls, and 10 ½ years in boys). This phase is under the control of growth hormone and sex hormones acting together.



ABNORMAL GROWTH IN CHARGE

- Failure of any of these 3 phases leads to short stature, and all of these are seen in children with CHARGE.
- Short stature is common in CHARGE, and about 75% have both height and weight at or below the lower end of the normal range.

FINAL HEIGHT

- Patients 19 years or older.
- Males (N=5) were 155 – 182 cm (5'1" - 6'):
Average 167.5 cm (5'6").
- Girls (N=6) were 150 – 170 cm (4'11" – 5'7"):
Average 161 cm (5'3").

Blake et al., 2005

SEVERE GH-DEFICIENCY

Severe growth hormone deficiency (GHD) is usually easy to diagnose. Affected patients are:

- Short
- Slowly growing
- Usually relatively overweight for their height (as growth hormone breaks down fat).
- In addition the skeletal maturation (the “bone age”) is delayed.

GHD & CHARGE

- Less severe forms of GHD are more difficult to diagnose.
- As a rule, however, the best predictor of whether a child has GHD is whether their growth rate is normal or not.
- There are, however, other reasons other than hormone deficiencies for poor growth in CHARGE

GROWTH HORMONE THERAPY IN CHARGE

Growth hormone databases

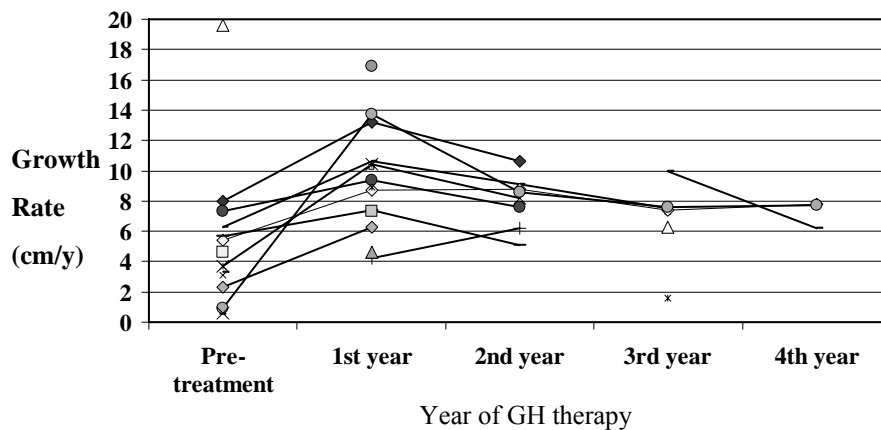
Data on GH therapy in CHARGE syndrome has been obtained from 2 large databases:

- National Cooperative Growth Study (NCGS: USA: (N=26))
- Kabi International Growth Study (KIGS: International: (N=32))
- Total of 58 patients (31 female)

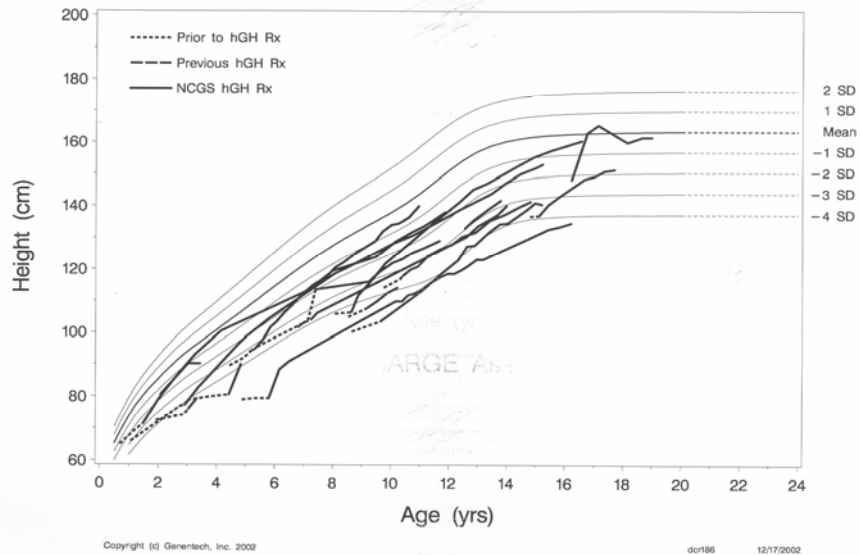
GH THERAPY IN CHARGE (NCGS)

- 26 patients (19 female) aged 0.5-16.7 years. All but one patient were pre-pubertal.
- Most patients were short (23 (88%) had heights below 2nd centile).
- 22 have had anterior pituitary function performed; 17 are GH-deficient.

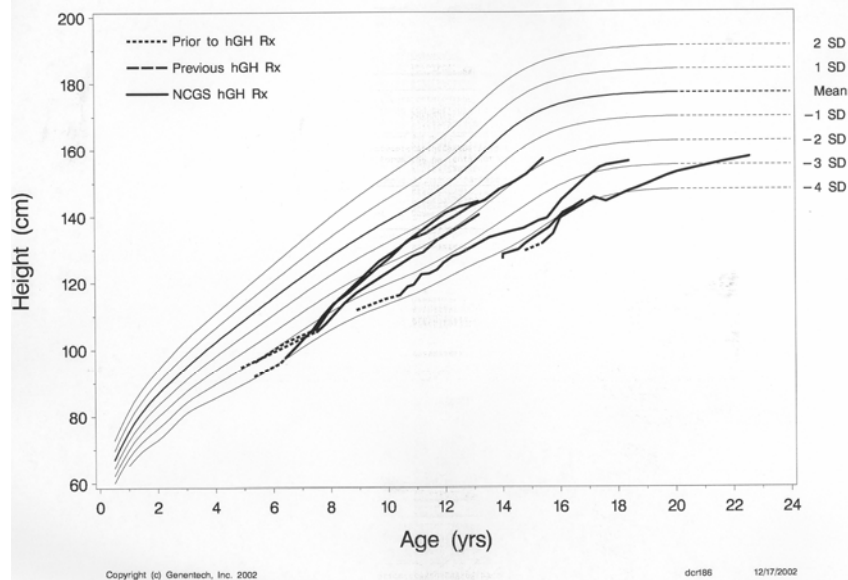
GH therapy in CHARGE (NCGS)



NCGS Patients with CHARGE Association – Females



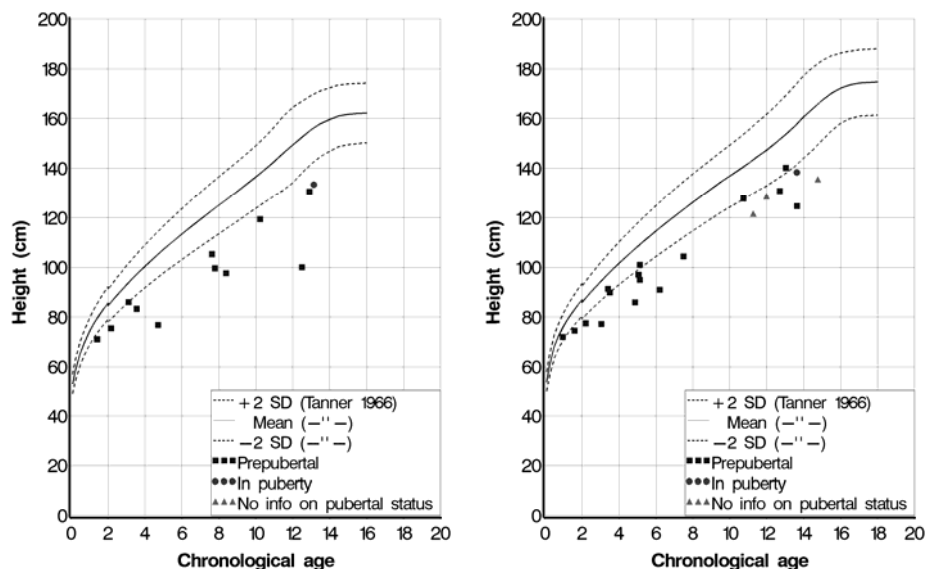
NCGS Patients with CHARGE Association – Males



GH THERAPY IN CHARGE (KIGS)

- 32 patients (12 female) average age 7.44 years. All but two (1 male, 1 female) were pre-pubertal.
- Most patients were short (27 (84%) had heights below 2nd centile)
- 26 have had anterior pituitary function performed. Mean GH level was 9.38 $\mu\text{g/L}$ (normal > 10).

CHARGE syndrome (KIGS): Baseline data Height at GH start. *Girls (left) & Boys (right)*



KIGS – CHARGE syndrome. Prepubertal longitudinal growth

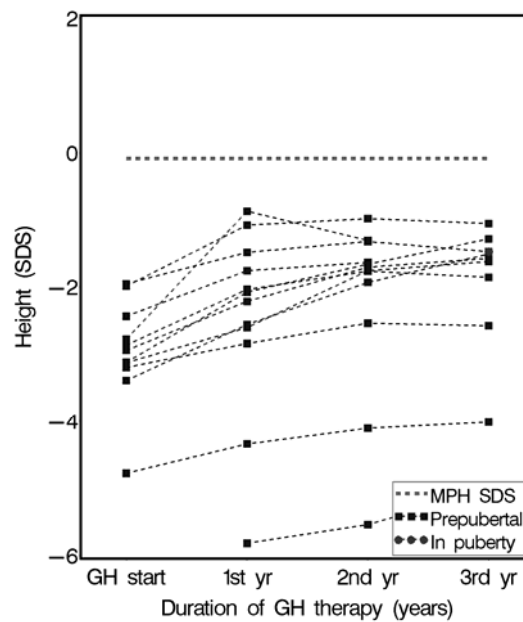
Variable	N	Median	10th pctl	90th pctl	Mean	SD	n(%)
Sex, boys	13						8(61.5)
Birth weight SDS	12	-0.85	-1.82	0.71	-0.62	1.10	
Birth length SDS	9	-0.22	-2.09	1.92	-0.24	1.08	
Mid parental ht SDS Tanner	13	-0.08	-0.92	1.21	0.09	1.04	
Max GH peak ug/L	11	6.90	5.50	8.20	8.61	6.85	

Height velocity (HV): KIGS

	All patients (No.)	Pre-pubertal patients (No.)
Pre-treatment HV (cm/y)	5.85 (N=9)	4.95 (N=5)
1st year HV		8.59 (N=13)
2nd year HV		6.82 (N=13)
Latest visit HV	5.14 (N=22)	

**CHARGE syndrome
Height SDS before and
during GH therapy**

**Patients with
2 years data (KIGS).**



Conclusions

- A number of patients with CHARGE have been treated with GH.
- Most were short, and, at least in the NCGS data, many were GH deficient.
- There is an improvement in growth initially on GH data.
- This does not appear to be sustained long-term, and final height data in these patients is lacking.

Conclusions

- Further analysis of the current data is required, to see whether GH deficient patients have a better response to therapy.
- Further data and studies are also required to see whether GH is of benefit in patients with CHARGE.



PROGRAM SECTION 26

Breakout Session #6
Saturday, July 28, 2007, 10:30-11:45
Pacific Ballroom III

Educational Implications
on Learning for a Child
with CHARGE



Panel Discussion Moderated by
Martha Majors and Sharon
Stelzer

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

26 ED Educational Implications on Learning for a Child with CHARGE (*Panel Discussion: Moderator: Martha Majors, Assistant Supervisor Deafblind Program Perkins School for the Blind and Educator/Teacher: Sharon Stelzer, Teacher Deafblind Program Perkins School for the Blind*)

Discussion will focus on a variety of teaching strategies that assist children with CHARGE to be more effective learners in the school and home settings. Specific strategies will be defined and examples given as to how to implement these strategies across all learning environments. These examples will be directly linked to the IEP content related to accommodations and specially designed instruction.

The poster session will be more specific and individualized for parents; the educators and parents will have time together to develop IEP content related to accommodations and specially designed instruction. The parent should be able to take home a worksheet that will assist in the development of the IEP.



PROGRAM SECTION 27

Breakout Session #6
Saturday, July 28, 2007, 1:00-2:00
Pacific Ballroom IV

Creating Community for
Adult Life: Planning an
Effective and Exciting
Transition



Presented by
Jackie Kenley and Nancy
Cornelius

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007



Creating Community for Adult Life:

*Planning an Effective
and Exciting Transition*

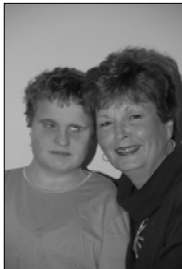
Presenters

Nancy Cornelius, Regional Representative
National Family Association for Deaf Blind

Jackie Kenley, Family Specialist
California Deaf-Blind Services

and most especially
**Mothers of daughters with
CHARGE**

Credentials



Mom to Laura Kenley, 22

- Happy, great smile and giggle, desire to communicate, loves chocolate! Loves to swim
- Profoundly deaf, light perception left eye, limited vision right eye
- Developmentally delayed
- Heart defects
- Seizure disorder

Credentials



Mom to Amy Cornelius, 21

- Bossy, funny, communicative, determined and resilient
- Profoundly deaf, low vision right eye only
- Developmentally delayed
- Heart defect
- Behavior issues
- G-tube fed
- Laryngeal spasms

What is Transition, Anyway?



We experience many transitions throughout life. In this session we are exploring the transition of young adults with CHARGE, from school to adult life.

Looking for OZ???



*Perhaps your vision is blurry
and
perhaps OZ looks
differently than you think!
Maybe more like Kansas?*

Planning for the Future



- It is essential to project into the future in order to create an effective transition plan.
- There are many futures planning tools to assist in identifying desired outcomes.

Planning Tools



- Its Never Too Early, Its Never Too Late!
- MAPS—Making Actions Plans
- Student-Family Focused Transition, Education and Planning

Just to name a few

It is never too early to begin.....



- Documentation
- Futures Plan
- Worksheets
- Videos- Communication and work resumes
- Local Resources

Strategies for Planning Effective Transition from School to Adult Life



- Know the Individuals with Disabilities Education Improvement Act. It is the federal law that mandates appropriate transition services and it drives local programs.
- Research local programs and get to know your community options.
- Know definitions and terms used by local service agencies.

Individuals with Disabilities Education Improvement Act

<http://idea.ed.gov>

The term "transition services" means a coordinated set of activities for a child with a disability that:

Is designed to be within a results-oriented process, that is focused on improving the academic and functional

achievement of the child with a disability to facilitate the child's movement from school to post-school activities, including postsecondary education, vocational education, integrated employment (including supported employment)

A Tale of Two Cities

- San Francisco
 - City College
 - Rec Center
 - Toolworks
 - Supported Employment through Adult Service Provider
 - Regional Center vendored services
- San Diego
 - State College
 - Community College
 - Trade School
 - Employment
 - Supported employment
 - Regional Center vendored services



Clear Communication is Essential!

I said...

- I would like to explore and observe some community placement **options** for my daughter.

I thought: supported living, group-home, or institution.

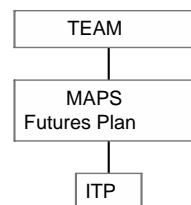
They heard...

- **Placement.**

They thought:
"placement" equals
group-home
placement.

No options about it.

Use Planning Tools and Your Child's Team



Who Knows and Works with Me??



Planning team

- Focus Person
- Family
- Friends
- School Team-Teacher, Aide, DIS Staff (therapists), Gen. Ed Teacher, Program Administrator, Other Specialists
- Adult Service Provider Rep
- Welcoming Agency Rep

Individual Support Needs

- Interpreter/Intervener
- Vision & Hearing Specialist
- Mobility Specialist
- Personal Attendant
- Job Coach
- Livings Skills Coach
- Behavior Specialist
- Medical Specialist



Adult Service Providers

- Habilitation/rehabilitation
- Local Day Programs
- The ARC
- State Department
- Federal Department
- Regional Center (CA)
- Transportation Services
- Services for the Blind
- Services for the Deaf
- Mental Health Services



How the Futures Plan Relates to the Student's Individual Education Plan & Individual Transition Plan

The futures plan identifies:

- Hopes and dreams of the focus-person
- Likes and dislikes
- Strengths upon which to build
- Weaknesses to be strengthened
- Personal and community links
- Healthcare, communication, and behavior support needs.

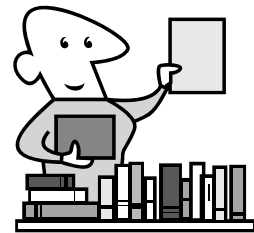
How the futures plan relates to the IEP/ITP

The preferences identified by the futures planning process are the basis for the goals and objectives on the Individual Transition Plan



Educational Options

- College
- Community College
- ROP
- Other training options



Vocational Options

- Paid employment
- Volunteer work
- Sheltered workshops
- Part-time/Full-time
- With or without support



Living Options

- Home with family
- On own with or without roommates
- Supported living
- Group home
- Institution



Recreation/Leisure Options

- Transportation
- Supervision
- Friends
- Activities/Events
- Communication



Getting from the Plan to the Reality

- Learn how to make lemons into lemonade
- Self-reflection and assessment
- Focus, creativity, flexibility, listen
- Identify local resources
- Get on waiting lists, if necessary
- Identify funding sources (SSI/SSA)



Making it Happen

It ***still*** takes a village!



Resources for Planning

- Texas School for the Blind-Transition from School to Adult Life in the Community -A *Two-part Planning Sheet*
<http://www.tsbvi.edu>
- *It's Never Too Early It's Never Too Late*
<http://www.mncdd.org/extra/publications.htm>
- *A Good Life*
<http://www.gifttool.com/shop>
- *How to Create a Winning Video Resume*
<http://www.sfsu.edu/~cadbs>

Resources, continued

- Suggestions for Creating Successful Transitions from School to Adulthood
<http://www.sfsu.edu>
- Transition to Adult Living and Information and Resource Guide
<http://www.calstat.org/info.html>
- Student-Family Focused Transition, Education, and Planning
Google-John R Johnson, PhD
- IDEA
idea.ed.gov
- D-B LINK
www.dblink.org search transition



PROGRAM SECTION 28

Breakout Session #7
Saturday, July 2, 2007, 2:15-3:15
Pacific Ballroom III

Inclusion: Using Person
Centered Planning
Strategies to Bring
Dreams to Reality



Presented by
Donna Consacro, Susan
Hiscutt, & Nancy Steele

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Inclusion: Using Person Centered Planning Strategies to Bring Dreams to Reality

8TH International CHARGE Syndrome Conference
Costa Mesa, California
July 28, 2007

Presenters:
Donna Consacro, Project TREDs
Susan Hiscutt, Parent
Nancy Steele, NCDB

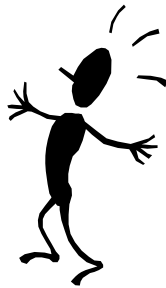
Thanks to Kathy McNulty, NCDB and Debbie Cate, TN DOE for materials used in this presentation.

What is Inclusion?

- Inclusion is a horizon for goal setting and problem solving....
John Obrien, Part of the Community
- True inclusion is [an] attitude that all students belong everywhere, with everyone else, in the school community.
Patrick Schwarz, From Disability to Possibility
- Full inclusion occurs when students with disabilities are full-time members of age-appropriate, regular classrooms in their home school (whatever school they would attend if they were not disabled), and receive any supports necessary to accomplish participation in both the learning and the social communities of their peers.
Lori Goetz, Including Deafblind Students: Report from a National Task Force

What is Person-Centered Planning?

A process that promotes positive outcomes!



Person-Centered Planning Is..

- an ongoing process
- a way to get to know the student and what he/she wants in life
- a means of understanding the dreams of family members
- a record of current events for future reflection
- the basis for meaningful educational and life planning
- voluntary and incorporates a collaborative philosophy
- brings to the table a variety of groups that play a role in the student's life

Person-Centered Planning...

- plans **WITH** not **FOR** the student (and family)
- starts with **abilities** and interests, rather than disabilities
- provides a comfortable atmosphere for the student and family
- hears from the student and family **FIRST**
- offers a better understanding of a family's dreams and fears
- give a visual and immediate picture of the situation
- provides helpful information to identify and prioritize needs

What it is NOT!

- Person-centered planning is not a "one-shot" deal. It's an ongoing process. Like life and transition itself, it's not static.
- It's not done with one or two people only.
- It's not done without the person's participation. As much as possible, the person participates in the meeting and is present at the meeting.
- It's not an IEP meeting, though it can contribute to developing IEP goals.
- It's not program centered. Its **person** centered.
- It's not a process that you learn in one training. You must be exposed again and again.

Pre-Planning for PCP

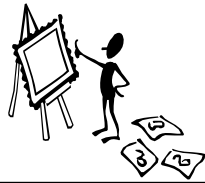
- Make the location family friendly
- Ask the family who they want to attend
- Give the family questions to think about before so they know what to expect
- Try to include the student as much as possible
- Determine who will coordinate logistics

What's In a Person Centered Plan?

- Person Centered Plans include sections for:
 - *Student and Family History/Profile*
 - *Student Interests & Abilities/Preferences*
 - *Student and Family's Hopes/Dreams*
 - *Student and Family's Fears/Concerns*
 - *Determining what's needed to turn dreams into reality and overcome fears*
 - *Development of an Action Plan*

Some PCP Tools

- **Personal Futures Planning**
- **Essential Life Planning**
- **PATH**
- **Routines-Based Interview**
- **MAPS**



Which PCP Tool Works Best?

"The choice of method is more like choosing a musical instrument than it is like selecting a hammer or a screwdriver from a toolbox..."

Marsha Forest, Jack Pearpoint & John O'Brien,
"What is Person Centered Planning", *All My Life's A Circle*.

MAPS McGill Action Planning System

- **History** – the whole nine yards
- **Dreams** – if wishes come true
- **Nightmares** – the worst fears
- **Who Is ___?** - short positive descriptors
- **Strengths/Gifts/Talents** – what I do well
- **Needs** – what I need / my team needs to help me do it
- **Modifications** – what makes life and learning accessible

Falvey, M.A., et al. *All My Life's a Circle: Using the Tools: Circles, MAPS, PATHS*

MORE MAPS

- **Choices Child Makes for Self** – practicing self-determination
- **Choices Adults Make for Child** – fostering self-determination
- **Preferences** - what works/what does not work
- **Perfect Day** – at this very time if everything was perfect
- **Priorities** – what are the most important things right now
- **Action Plan** – what, who, when, how

Falvey, M.A., et al. *All My Life's a Circle: Using the Tools: Circles, MAPS, PATHS*

MORE MAPS

- **Communication Maps** (Family, Friends, Service Providers)
 - *Expressive: Who the child communicates with and how the child communicates (expressive)*
 - *Receptive: Who communicates with the child and how they communication*
- **Relationship Maps** (Family, Friends, Service Providers)
 - *Who does this child come into contact with: family, friends/community (free), service provider (paid)*
- **Places Maps**
 - *Where a child goes in the home, school, and community*

A MAPPING EXAMPLE

Aaron's Story!

Aaron's History

Born July 23, 1997
Six days later, learned about
CHARGE Syndrome

Aaron's History

*TEIS started 10 days after birth *OMT started
at 6 weeks *PT started at 2 months *TIPS
started at 9 months *ST started at 10 months
*OT started at 30 months

Aaron's History

*Feeding Issues
First 5 months bottle feed, OG tube feed for 7
months Had G-tube surgery in 1998
Still uses G-tube, but is now eating food

Aaron's History

*Hearing Issues
Born with profound to severe hearing loss
Got 1st hearing aids at 10 months
In late 2000 got FM system
In July 2002 got new hearing aids

*Vision Issues
Bilateral colobomas of the optic nerve
Very limited vision in right eye, limited vision in left eye
Aaron's functional vision is very different from what
assessment says

Aaron's History

Aaron attended ECLC from 22 months to 3 years old.
Couldn't sit up yet
Made progress after watching the other students!
At 3 years old, Aaron went to Gunnings Special School

Aaron's History

Returned to ECLC at 4 years old.
In 2002 Aaron began Rock Springs Elementary
And in 2003 Aaron started Kindergarten at Miller Perry
Elementary School and is currently in 4th Grade at
Miller Perry.

Aaron's Dreams

To be safe
To have friends
To read

Aaron's Dreams

To go to a restaurant and order his own food
To live on his own To be happy
To be able to talk
To be an active community member

Aaron's Dreams

To have more choices
To be happy!
To graduate from college
To hear as much as possible
To have a sweetie
To change the world

Aaron's Nightmares

- ✗ Being picked on or neglected
- ✗ Being taken advantage of
- ✗ Not being able to eat on his own
- ✗ Not meeting all his dreams
- ✗ Being underestimated
- ✗ Poor communication skills
- ✗ Being frustrated & giving up
- ✗ Medical needs aren't met

Who is Aaron?

*More patient *Fighter *Persistent *Curious
*Socially Selective *Handling change much better
*Perfectionist *Unique *Equestrian *Watcher
*Stubborn *Independent *Adaptable

Who is Aaron?

*Expressive *Happy *Regimented *Manipulator
*Explorer *Cautious *Resourceful *Kind
*Determined *Performer *Funny *Intelligent
*Creative *Cute *Communicator (in his own way)
*Loved and loving *Respectful of others' space

Aaron's Strengths

*Compensation Skills
*Stubborn, strong willed and self-determined *Curious
*Great at puzzles *Computers
*Building things *Problem-solving *Horseback riding
*Manipulates mouse
*Winning people over with charm *Inspiration *Ability to continue improving
*Support system *Open to opportunity *Memory
*Understanding space

Aaron's Perfect Day

To sleep all night in my own bed
To wake up happy, by myself
To communicate with others
To be in a regular ed class
To use the general curriculum
To socialize with my friends
To have lunch in the cafeteria
To attend all related arts classes

Aaron's Perfect Day

To learn something every day
Therapists help modify
Therapy will be after school
Teachers would have a perfect day too!
I will go to McDonalds after school, play sports, get dirty and do my homework.
Dinner with Mom & Dad
Go to bed by myself

Aaron's Needs

*To be understood *Have a communication system
*Strategies to develop peer relationships *Continued high expectations
*Inclusion *Strategies for peers
*To be challenged *Maximize therapy resources *Better communication among team members *Daily schedule
*Choices *Team that thinks outside the box *Continued auditory development
*Team sharing new information
*Aaron eating more independently
*Embedded therapies

We Have Great Information About Aaron— How Do You Put It to Good Use??

An Exemplary Program

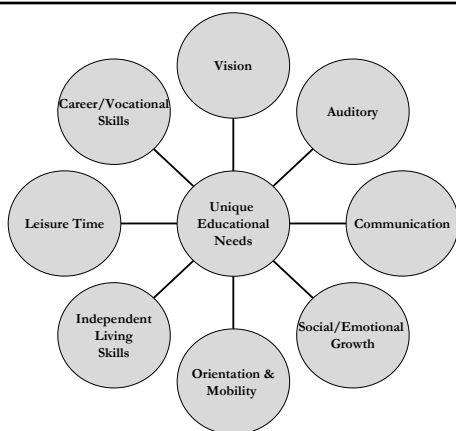
- Uses a collaborative team approach to developing IEP goals and objectives based on student's needs.
- Uses a collaborative team approach to implement the educational program
- Curriculum is functional, activity-based age-appropriate and uses schedules, routines and natural environments

Exemplary Programming

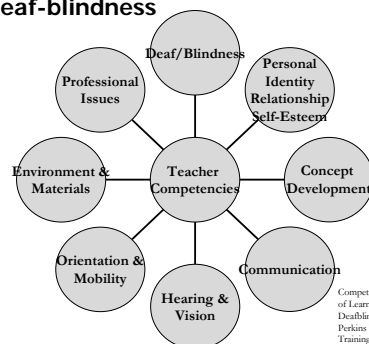
- Uses a collaborative team approach to evaluate student outcomes
- Team members share the responsibility for collecting data on student progress towards IEP goals

Exemplary Programming

- School administration supports the collaborative team approach to providing time for the team meetings and planning
- School personnel maintain high quality instruction
- High level of coordination between special and general education
- Involvement and progress in the general education curriculum
- Participation in regular education settings.

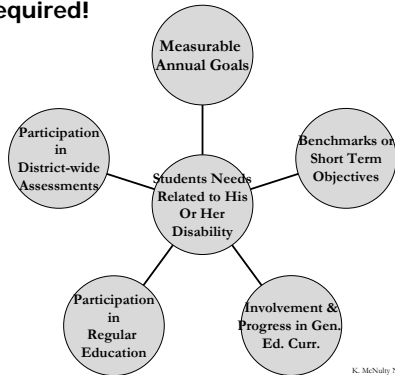


Teacher Competencies in Deaf-blindness



Competencies for Teachers of Learners Who are Deafblind
Perkins National Deafblind Training Project
B. McLetchie M. Riggio

A Collaborative Team Approach Required!



K. McNulty NTAC, 2002

Challenges to the Collaborative Process

- Resource Limitations
- Professional Autonomy
- Meetings Can Be Risky Business (Kieffer, 1988)
- TIME!

Critical Elements of Collaborative Teaming

SHARED.....

NEED	EXPERTISE
VALUES	LEADERSHIP
GOALS	OWNERSHIP
TEAM CULTURE	

Collaborative Teaming & A Shared Need

- Students who are deaf-blind need specialized educational strategies and interventions to ensure productive, fulfilling lives.
- Personnel who work with a student who is deaf-blind should have a clear understanding of the effects of these disabilities on a home & school environment

Collaborative Teaming & Common Goals

The educational program-

- ✓ Builds a framework
- ✓ Provides common goals
- ✓ Clarifies expectations

COLLABORATIVE TEAMING
 +
SHARED OWNERSHIP =
EXPERTISE

Characteristics

- All stakeholders represented on team
- Uses a collaborative teaming process that involves:
 - **Face to face interaction**
 - **Positive interdependence**
 - **Individual accountability**

Characteristics

- All members have unique & needed expertise
- Equal valuation of each member's input
- Members alternately engage in dual roles of:
 - » Teacher & learner
 - » Expert & recipient
 - » Consultant & consultee

Collaborative Teaming & Shared Leadership

Functions of the traditional leader are distributed among the group

Team Culture

How the Team Conducts Its Business

- ❖ Face to face interaction
- ❖ "We're all in this together!" feeling
- ❖ Focus on small group interpersonal skills that develop trust, effective communication and leadership
- ❖ Focus on developing skills in creative problem solving, decision making and conflict management

Typical Team Problems

- Lack of Common or Clear Goals
- Poorly Defined Membership or Roles
- Ineffective Communication Processes
- Ineffective Meetings Processes

GOLDEN RULE:

**Goals – Roles –
Process**

**There is a PROCESS to
running a good meeting**

#1 BE ORGANIZED.

- ✓ Know the goal of the meeting
- ✓ Make sure members know their roles
- ✓ Use an agenda
- ✓ Set timelines in advance

**#2 ENCOURAGE
PARTICIPATION**

- Ask questions
- Test for understanding
- Strive for consensus
- Bring in the quiet members
- Minimize the monopolizers

**#3 RESOLVE ISSUES –
ACTION PLAN**

- ❖ Work on problems and issues
- ❖ Develop an action plan
- ❖ Everyone gets a task

**#4 SUMMARIZE AND
WRAP UP**

- ✓ Make sure every agenda item was discussed
- ✓ Review action plan
- ✓ Make sure everyone knows next steps
- ✓ Set date for next meeting

**SAMPLE GROUNDRULES
FOR A MEETING**

- ✓ Honesty
- ✓ Don't attack
- ✓ Don't interrupt when another is speaking
- ✓ All views are valid - even if we disagree
- ✓ No side chats during the meeting
- ✓ Respect feelings
- ✓ Ask questions for clarification
- ✓ Don't monopolize the conversation

Aaron and Inclusion

Inclusion at Home

*Very easy for our family to do
*Grandparents, parents, aunts, uncles, cousins learning sign language
*Family has always treated Aaron as "normal"
*Always included in family functions

Inclusion at School

*Attends Regular Ed classes *Has a one-on-one aide
*Therapies are embedded *Only "pulled-out" as needed
*General Ed Curriculum is adapted and modified

Inclusion in the Community

*Plays t-ball in community league
*Member of a local Cub Scout Den
*Attends church with Mom & Dad
*Goes Bowling & Putt Putt
*Eats out at local restaurants
*Loves to go shopping with Dad

Helpful Suggestions

- If possible introduce student to participants he/she does not know before the meeting.
- Limit number of persons attending meeting to top priority participants.
- Bring maps from previously held PCP meetings to use as examples or work on student's map prior to the PCP meeting.

Key Team Members

- The Family
- The School
- The Community
- Additional Supports
 - TREDs (Tennessee's Deafblind Project)
 - NCDB (National Consortium on Deaf-Blindness)

Key Strategies for Support

- Start Early
- Identify Key Members
- Team Building
- Needs Assessment
- Communicate/Team Meetings
- Think Outside the Box
- Share Individual's Expertise
- Training followed by in class and home support

Benefits of MAPPING

- Creates a complete and holistic picture of the student
- All participants share common knowledge about the student, gain insight from others
- Looking at the MAPS begins to suggest goals to work on and skills to build
- Planning is based on preferences and interests
- Wisdom of the group is greater than any one individual—Creative ideas emerge!

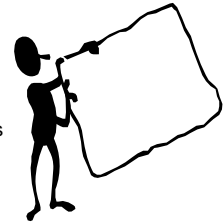
Criteria for Success

- » Shared values
- » Ability to see capacities & strengths
- » Attitude of discovery
- » Commitment to follow-up
- » Comfort with small steps
- » Perseverance
- » Ability to brainstorm
- » Good facilitator

K. McNulty, NTAC, 2003

PCP Resources

- NCDB
- State DB Project
- DB-LINK
- State Inclusion Projects
- State Transition Projects
- Parent Training Centers



Resources

- Websites for further information:
 - **Person Centered Planning & Inclusion**
 - Cornell University Employment and Disability Institute, www.ilr.cornell.edu/ped/tsal/pcp/index.html
 - Pacer Center, www.pacer.org/tatra/personal.html
 - Routine Based Interview Resources, www.vanderbiltchildrens.com/interior.php?mid=1173
 - MAPS & PATH Resources www.inclusion.com
 - American Association of Intellectual and Developmental Disabilities http://www.aamr.org/Policies/faq_planning.shtml
 - National Center for Children & Youth with Disabilities, Planning for Inclusion, www.nichcy.org
 - Power of 2, www.powerof2.org
 - Kids Together, Inc., www.kidstogether.org
 - Circle of Inclusion, www.circleofinclusion.org
 - Personal Futures Planning, Beth Mount, www.capacityworks.com/books.html

Questions & Answers



References

- Brown, J.G & Haynes, D. (2000) **Reach for the Stars: Planning for the Future**. Louisville, KY: American Printing House for the Blind.
- Falvey, M.A., Forest, M., Perpoint, J., Rosenberg, R.L. (2000) **All My Life's a Circle: Using the Tools: Circles, MAPS, PATHS**. Toronto, Canada: Inclusion Press.
- Goetz, L. (1997) **Including Deafblind Students: Report From a National Task Force**. San Francisco, CA: San Francisco State University.
- McWilliam, R. A. (1992) **Family-centered intervention planning: A routines-based approach**. Tucson, AZ: Communication Skill Builders.

References (continued)

- Mount, B. (1990) **Making Futures Happen**. Available on the web at http://www.mncdd.org/extra/publications/Making_Futures_Happen.PDF
- McWilliam, R.A. & Clingenpeel, B. (2003) **Functional Intervention Planning: The Routines-Based Interview**. Center for Child Development, Vanderbilt University. Nashville, TN: www.vanderbiltchildrens.com/interior.php?mid=1173
- Nisbet, J. & Hagner, D, eds., (2000) **Part of the Community: Strategies for Including Everyone**. Baltimore, MD: Paul H. Brookes Publishing Co.
- Pearpoint, J., et al. (2001) **PATH: A Workbook for Planning Positive Possible Futures**; Inclusion Press
- Schwarz, P. (2006) **From Disability to Possibility: The Power of Inclusive Classrooms**. Portsmouth, NH: Heinemann.

References (continued)

- Smull, M. W. (2007) An Overview of Essential Lifestyle Planning (ELP). Available at <http://www.ilr.cornell.edu/edi/pcp/course05b.html>
- USARC/PACE & Allen, Shea & Associates. (2000). Listen to Me! Available at <http://www.learningcommunity.us/documents/ListenToMe.pdf>



PROGRAM SECTION 29

Breakout Session #7
Saturday, July 28, 2007, 2:15-3:15
Pacific Ballroom IV

Pain Issues in CHARGE



Presented by
Veronika Bernstein

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

Pain Issues in CHARGE

Veronika Bernstein, Ph.D.
Developmental Specialist

Perkins School for the Blind

Veronika.Bernstein@Perkins.org

Introduction

- Good Afternoon
- My background is in brain-behavior. I have been working at Perkins for about 20 years and in the field for about 35 years
- I would like to learn something about you:
 - How many children do you have or do you work with?
 - How old is your child with CHARGE?
 - Does your child have “out of the blue”, “don’t know why” episodes?
- Please DO INTERRUPT me with remarks, questions and answers

Painful Questions

- How many of typically developing children and adults have chronic or recurring pain?
- What does it mean that children with CHARGE have an “increased pain threshold”?
- Do children and adults with CHARGE receive adequate treatment for pain?
- How to recognize pain in your child?
- How to help the child to cope with pain?

Pain Facts

- Pediatric chronic pain affects 15-20% of typically developing children
- Acute pain in children is managed more effectively as compared to chronic pain
- Adults: 9 out of 10 Americans suffered from regular pain in the past 6 months
- Pain Management: 43% of adults report good pain control, the rest are living with recurring moderate to severe pain

Pain and Developmental Disabilities

- Years of debates by professionals whether people with developmental disabilities experience pain (parents always knew)
- People with developmental disabilities have been excluded from research studies
- Expression of pain in the absence of words
- Subjective recognition of pain by caregivers

Pain. The Concept

- Pain is a multidimensional experience that includes sensation, emotion, thoughts, and feelings.
- Pain is always a subjective experience. As such, pain cannot be observed directly but must be inferred and interpreted by others.

Pain Mechanisms

- Peripheral: muscles, joints, bones
 - Different conduction velocities,
 - A-beta, A-delta and C fibers
- Motor:
 - Spinal cord
 - Complex system of transmission
- Emotional:
 - Thalamus
 - Relay stations of emotional processing
- Cognitive:
 - Cortex
 - Multiple areas and complex processing

Physiological Pain Response in People with Developmental Delay

- 1. General decrease in sympathetic modulation in people with developmental delays (heart rate, blood flow, neuro-hormonal changes) - it takes longer to activate as well as it takes longer to stabilize
- 2. Physiological response to pain may be reduced or qualitatively different similarly to other (environmental) stimulation - it takes longer to perceive, to process, to act on
- 3. Early pain experience alters physiological processing of the pain

Acute Pain

- Response to a specific cause
- Time limited
- Stops in response to body healing
- Stops in response to treatment
- Examples: vaccination shots, sore muscles after workout, accidental bumps & cuts

Chronic or Recurring pain

- A non-specific response to often unknown cause
- Long term (weeks, months, years)
- Persistent
- Does not stop after treatment of the initial cause
- Examples: back pain, organ pain (visceral), phantom pain, neuralgias, migraines

When Pain is in the Brain

- Pain receptors get stuck in an "ON" mode
- Long term memories of painful experiences surface again and again in response to some environmental cues
- Pain in the brain is not a symptom of a problem but a disease itself
- Hormones affect pain receptor sensitivity (menstrual cycle, developmental hormonal changes e.g.)
- Cultural aspects of pain

Chronic Pain in Adults

- Absence of clinical research in people with developmental disabilities
- Typically developing adults:
 - Strong link between chronic pain and anxiety
 - PTSD, stress, glucocorticoids and chronic pain
 - Amygdala, fear response, depression
 - Hippocampus, pituitary-adrenocorticals
 - Hormones
 - Belief Systems

Neurological Impact of Early Pain

- Animal Models:
 - Neonatal pain leads to structural and functional reorganization of pain system (peripheral and central)
 - Decreased pain perception locally
 - Repeated low pain leads to decreased central perception
 - Increase in pain inhibitory systems

Human Infants

- Pain can lead to increased or decreased cerebral blood flow
- Painful/stressful events cause fluctuations in intracranial pressure leading to hemorrhage and other conditions
- Tactile threshold is significantly lower at lower gestational ages as well as habituation
- Neonatal pain has long lasting effects on arousal regulation

Long-term Effects of Early Pain

- Retrospective studies of children with severe cognitive impairments point to increased self-injury
- Prospective studies do not support this
- Children who have repeated admissions and multiple surgeries show decreased language functioning
- One study (Ludman et al, 2001) found continued impact of early pain as late as at 11-13 y.o.

How we COPE with Pain

- Increased and / or Decreased Sensitivity to pain takes place on some or all of the four levels:
 - 1. Peripheral (muscles, joints, bones)
 - 2. Motor (Spinal)
 - 3. Emotional (Thalamic)
 - 4. Cognitive (Cerebral)

Pain Assessments

- 1. You know when you see it. You yourself have experienced similar pains in the past. You interpret your observations: "My child is in pain"
- 2. Medically based rating scales for acute pain (surgery e.g.)
- 3. A pragmatic mix of #1, #2, and behavioral interventions to help child to cope with pain - by Veronika, here and now.

Pain Rating Scales - 1 (refer to handouts)

- "Faces Pain Scale"
 - Five or six simple cartoon faces
 - Emotionally neutral expression on the left
 - Distressed and grimacing face on the right
 - The child is asked which face best represents how much pain there is
 - Requires good visual and cognitive skills

Pain Rating Scales -2 (refer to handouts)

- Visual analogue scales
 - “No Hurt” on the left “Worst Hurt” on the right
 - Numbers 0 to 5
 - Pain thermometer; vertical representation
 - The child is asked to point to the place on the line that best represents the level of pain

Pain Rating Scales - 3 (refer to handouts)

- Pediatric Pain Questionnaire (PPQ):
 - Multidimensional questionnaire
 - Assesses the child's participation in activities
 - Level of participation
 - How the pain has affected the child's emotional state and relationships
 - There are versions of PPQ for older children and adolescents

Pain Rating Scales -4 (refer to handouts)

- NCCCPC-R Non-communicating Children's Pain Checklist - Revised:
 - Designed specifically to help parents and caregivers identify clues that may indicate pain
 - Autistic children
 - Neurological impairments
 - Severe brain injury
 - It is behavioral observation scale
 - See the summer issue of CHARGE Accounts 7/07

Pain Rating Scales -5 (refer to handouts)

- Faces, Legs, Activity, Cry, Consolability (FLACC):
 - Behavioral observation checklist
 - Developed for ages two to seven
 - Used after surgery
 - Used during medical procedures

CHARGE Pain and Coping with Pain (Veronika)

- Survey of 23 children with CHARGE, March-July 2007)
- Age: 3 to 24
- Individual list of Behaviors during Chronic and Acute pain episodes
- Individual list of strategies (including medication) that HAVE NOT HELPED in the past
- Individual list of strategies (including medication) that HAVE BEEN HELPFUL in the past.

Pain Coping Strategies: Medical

- Analgesics (opiates controversy)
- Topical Pain Relievers
- Muscle Relaxants
- Antidepressants
- SSRI
- Injection or Nerve Blocks

Pills or Shots

- Most children would rather have pain than an injection
- Less than 1% of children treated with opioids develop addiction

Common Opioid Side Effects

- Nausea
- Vomiting
- Constipation
- Sedation
- Irritability

Pain Coping Strategies: Non-Medical

- Non-pharmaceutical strategies are currently recommended in most medical settings
- Non-drug coping strategies
 - Enhance comfort
 - Increase quality of life
 - Promote sleep

Pain Coping Strategies: 1. Peripheral

- Warm blanket
- Cold pack
- Touch
- Stroking
- Caressing
- Massage

Pain Coping Strategies: 1. Peripheral (cont.)

- Massage of the painful area or other areas:
 - Relieves pain
 - Relieves muscle spasm
 - Mobilizes contracted muscles
 - Increases blood flow and circulation

Pain Coping Strategies: 2. Motor

- Positioning:
 - Swaddle to limit excessive movements
 - Nestle by tucking around sides / back / feet
 - Assist in better body alignment (pillow between the knees e.g.)
- Rhythmic movement:
 - Rocking (swings, rocking chair)
 - Walking
 - Bouncing

Pain Coping Strategies: 2. Motor (cont.)

- Positioning Devices
- Corset
- Weight belt
- Orthopedic devices
- Wrist guards

Pain Coping Strategies: 3&4. Emotional&Cognitive

- Environment:
 - Shade eyes
 - Reduce lights
 - Reduce all noise
 - Turn off the phone
 - Cluster disturbing activities
 - Allow periods of undisturbed rest
 - Modulate the rate of activities (quick or slow)

Pain Coping Strategies: 3&4. Emotional&Cognitive

- Redirection and Distraction
 - Offer food
 - Allow fasting
 - Toys
 - Music
 - Video, TV
 - Other activities

Pain Coping Strategies: other strategies:

- Herbs
- Herbal creams and ointments
- Acupressure
- Reflexology points
- Chiropractic and osteopathic manipulation
- Yoga breathing and poses
- Strengthening exercises
- Flexibility exercises
- Hypnosis

Pain Coping Strategies: Summary

- We need to educate parents and care givers about the lasting effects of an early pain
- Have Individual Assessment for acute and recurring pain for your child
- Write Individual Pain Coping PLAN for your child
- Data to evaluate effectiveness of the pain coping plan

Child's Name _____

CONFIDENTIAL

PAIN AND COPING WITH PAIN

Age: _____

Date: _____

Information Provided By: _____

1. Recurring or Chronic Pain (abdominal colic, migraine, e.g.)

How often does the child have **Chronic** Pain?

Daily____ Weekly____ Monthly____ Rarely____ Never____

How often has the child have had **Chronic** Pain in the past?

Daily____ Weekly____ Monthly____ Rarely____ Never____

What is the nature of the recent **Chronic** Pain?

What was the nature of the **Chronic** Pain in the past?

What is the typical range of duration of **Chronic** Pain episodes (check all that applies)?

10-30 min.____ couple of hours ____ 6-12 hrs____ 12-24 hrs ____

on and off for several days____ other _____

2. Acute Pain (acute ear ache, cuts, bruises, medical procedures, e.g.):

1. How often has the child been in **Acute** Pain in the past six months?

Daily____ Weekly____ Monthly____ Rarely____ Date of Last Incident: _____

2 What was the nature of the **Acute** Pain?_____

Child's Name _____

CONFIDENTIAL

IDENTIFICATION OF PAIN EPISODE

Child can describe the pain with words like _____

____by signing ____in writing ____by drawings

Vocal: ____cries (with or without tears) ____moans ____screams / yells

____more persistent cry ____more high-pitched cry

other: _____

Facial: ____looks distressed ____eyes wide open ____eyes squeezed up ____eyes dulled

____mouth open ____frowns ____clenched jaw ____quivering chin

____lips pouting, tight, or puckering

other: _____

Physical: ____flinches from contact ____tense and stiff ____floppy ____kicking

____ legs drawn up ____arched back ____jerking ____itching ____agitated

____appetite (increased or decreased) ____sleep (increased or decreased)

other: _____

Physiological: ____breath holding ____gasping ____shivering

____skin flushed ____skin pale ____nausea

other: _____

Emotional: ____irritable ____angry other: _____

Social: ____withdrawn ____disinterested in favorite activities

____seeks to be cuddled ____resists being cuddled ____unable to be comforted

other: _____

Pain Location: ____points to ____rubs ____hits self ____protects or favors

Child's Name _____

CONFIDENTIAL

other: _____

STRATEGIES THAT HAVE NOT HELPED IN THE PAST:

Please list **medications that HAVE NOT HELPED** in the past and the reasons why:

STRATEGIES THAT HAVE HELPED IN THE PAST:

Environment: ____shade eyes ____cover with blanket ____avoid loud noise

____ undisturbed rest ____warmth ____cold

other: _____

Social: ____touch ____stroking ____cuddling ____massaging ____rhythmic rocking

other: _____

Successful Redirection or Distraction (please be specific):

Food: _____

Toys: _____

Music;: _____

Other Auditory: _____

Video, TV: _____

Other Visual: _____

Other Activities: _____

Child's Name _____

CONFIDENTIAL

MEDICATIONS THAT HAVE HELPED:

Any Additional Information:



PROGRAM SECTION 30

Breakout Session #8
Saturday, July 28, 2007, 3:45-4:45
Pacific Ballroom IV

A Neurodevelopmental
Perspective of CHARGE
Syndrome:
Impacts on Learning and
Behavior



Presented by
Kim Lauger

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

CHARGE Syndrome

A Neurodevelopmental Perspective

The impact of altered development on learning, behavior, and being

Kimberly Lauger, 2007

From early in gestation the neurodevelopment of an individual with CHARGE Syndrome is altered. When disruption of sensory systems occurs in utero it also impacts the neural pathways connecting those systems with the brain. At birth, medical crises and procedures, eating challenges, sensory impairments, balance problems and low tone continue to alter the developmental course. Now information coming into the brain must be organized and help mature the neural connections going to the cortex – the thinking brain. (Eliot 1999) This development is dependent on movement and experience, again altered with CHARGE.

With CHARGE, the most basic and essential tasks of life, breathing, eating, and moving are often impacted from the beginning. These stressors then alter the functioning of the autonomic nervous system, often priming it to respond quickly and fully to even minor stimulus. Due to the interconnectedness of systems, irregularities in one system impact the development of other systems.

An example is sucking. Sucking helps the two hemispheres of the brain work together; it helps the eyes come in and begin to develop the ability to focus up close; the eyes being able to focus helps the ears know what to focus on and what to ignore; sucking strengthens the sphincter muscle of the mouth, which also strengthens the other sphincter muscles, such as those which help the eye respond to light, and to those in the GI tract which control reflux and elimination. There is even some suggestion that sucking after birth lets the brain know I am out now and have more room to grow, stimulating the production of growth hormone. (Bluestone, 2004) Clearly this is a significant task developmentally. How does the interruption of sucking influence later developmental tasks such as being able to listen to words and block out environmental noises?

As bleak as the early developmental course in CHARGE may seem, it is important to remember that neurodevelopment occurs throughout the lifespan, with the brain in a constant state of adaptation based on the environment. One only has to watch individuals with CHARGE (admirably referred to as CHARGErs), individuals who in spite of incredible challenges show us what they CAN do in spite of these developmental changes. In fact, if proof were ever wanted for neural plasticity, the ability of the brain to figure out a new way when there is a disruption in the typical pathway, one simply need to watch a CHARGEr with impaired vision, hearing, and balance systems get up and walk. Generally in neuroscience one looks to periods of critical time periods for developmental skills to be learned. (Eliot 1999) CHARGErs also give proof that these are in truth optimal periods, not critical. (Bluestone, 2004) Yes, perhaps it would have been more efficient if the child had learned to walk or talk along typical timetables, but failure to do so then, does not necessarily prevent them from learning to do so later.

Yet, even with these incredible accomplishments and the fortitude shown by those with CHARGE, there is much discussion about difficulties in certain areas of functioning that persist. These difficulties or CHARGE quirks can cause problems with academic, social, or behavioral learning. A few of these concerns are listed below:

- Pain sensation; seeming insensitive to some pain and overly sensitive to other pain
- Complaints with hair washing, hair cuts, cutting finger nails
- Sleep disturbances, night parties, needing someone with them, trouble getting to sleep, waking too early
- A need for rules, order and routine, a need for others to follow these rules, but sometimes a hard time following the rules themselves
- Hand flapping, spinning in circles, and other self-stimulatory behaviors
- Difficulty following instructions
- Difficulty paying attention
- Communication
- Autistic-like behaviors; stimming, rituals, obsessions, acting on people to communicate rather than using words/signs
- Visual processing, binocular functions, light sensitivity
- Walking and balance issues
- Adjusting to change
- Needing deep input, liking hugs, disliking kisses
- Fatigue, being interested but not being able to participate long, how hard they work, how easily overwhelmed
- Eating difficulties, plus anxiety with eating
- Hearing sensitivities
- Underestimation of visual or auditory impact due to incredible coping strategies
- Melt downs or flare ups
- Math skills and handwriting

It is interesting to note that these quirks, behaviors that don't seem to make sense given how well functioning occurs in other areas, seem to show up in varying degrees in many people with CHARGE even though their developmental and life paths are so diverse. Depending on the severity of these difficulties, the individual with CHARGE may end up with additional labels, such as Autism, ADHD, or Obsessive Compulsive Disorder to name a few. Yet, these labels explain what we see, not why we see them and efforts are made to suppress the symptoms, rather than to address underlying causes.

In behavioral science we often look to antecedents to figure out what might precede these difficulties. A neurodevelopmental perspective adds a layer of considering how these antecedents are impacting this person based on the strengths and glitches in their neurodevelopmental systems. Attention is paid to how well these systems work independently and interdependently. Attention is paid to the efficiency of these systems and the amount of energy needed to perform the tasks. Understanding is gained at a new level, where the team can now figure out ways to 1) enhance that person's weak systems,

2) improved the efficiency between systems, 3) adapt the environment or expectation to alleviate physiological stress responses, and to 4) set up the environment and expectations such that the individual CAN perform differently.

The best examples I have found to explain a variety of possibilities for similar behaviors based on a neurodevelopmental model come from Judith Bluestone of the HANDLE Institute. Turning Point: Some Pivotal Insights Into Behavior and Learning is attached and is useful to share with educational teams.

For those wanting to know how neurodevelopmental systems develop before and after birth read What's Going On in There.

In addition, I highly recommend interested persons read The Fabric of Autism for further understanding of how systems impact function. Although the title says Autism, the examples regarding neurodevelopmental systems apply to all persons, particularly those with sensory sensitivities, and autonomic nervous system irregularities, as often seen in CHARGE.

For further information contact:

Kimberly Lauger

520-907-2842

klauger@comcast.net

References:

Bluestone, Judith, The Fabric of Autism, Weaving the Threads Into A Cogent Theory, 2004, The HANDLE Institute, Seattle, WA.

Eliot, Lise, What's Going On In There? How the Brain and Mind Develop in the First Five Years of Life, 1999, Bantam Books, New York, New York.

Glossary of Terms

Neurodevelopment – the development of the nervous system along the life span – prenatal to death

Neural Plasticity – the ability of the brain to adapt in response to the environment

Optimal Period – the optimal period for a task of neurodevelopment

Neurodevelopmental Systems – those systems that interact to influence the functioning of the nervous systems

System Definitions from Marianne Wagner, HANDLE of Olympia
Used with permission

Vestibular Functions - The vestibular system is involved in maintaining balance and experiencing gravitational forces, but also supports the processing of all other sense

impressions related to movement, including visual and auditory processing. It also plays a role in regulating attention. Stimulating the vestibular and tactile systems helps in processing information on higher levels, such as academic work.

Muscle Tone - Muscle Tone refers to the tension in the resting muscle. It affects how fast a muscle can respond to a stimulus, how much energy it takes, and how well it can regulate the response.

Proprioception (Awareness of body in relation to space) - Proprioception allows us to not only feel our own body's position, but also to sense our body in relation to space, and to experience space in relation to our body as a fixed point, including an awareness of direction. Amongst other, basic functions, this sense is involved in such learning processes as writing, reading and math and is important for being comfortable in crowds.

Sense of Touch (Tactility) - The sense of touch allows us to feel the quality of objects in our surroundings that we are touching with our body. Beyond that, the sense of touch also registers processes within our body. In that way, it has an important role in monitoring our inner environment and well-being. The sense of touch is involved in the processing of temperature and pain. The proprioceptive (tactile) receptors in the joints and muscles provide the proprioceptive system with information about the body's position.

Audition/Auditory Processing - Audition involves more than hearing well in both ears. Besides hearing the full range of sounds that the human ear can perceive, the brain also needs to assemble these sounds into meaningful words, music or other environmental sounds. The impressions received through hearing need to be coordinated with other impressions, for example sight and touch. There is an intricate interplay between the functions of the eyes and the ears and where the brain is to direct its attention. If this interplay is disrupted, it becomes difficult for a person to pay attention to the appropriate stimuli.

Visual Functions - Besides the ability to see clearly, the eyes need to be able to adjust their focus quickly to different distances, move smoothly in coordination and work as a team. The brain needs to be able to process the different information coming from both eyes in such a way that true binocular vision becomes possible.

Differentiation - This refers to the ability to separate out body parts, engaging the ones needed for a certain activity and keeping the others uninvolved.

Lateralization - Laterality is important in order to establish strong motor and visual skills as well as efficient higher cognitive functions. Besides having a dominant hand, eye, ear and foot, we also have specific areas in one or the other side of the brain that specialize in processing certain stimuli. Many people show processing and response patterns that are often attributed to one or the other hemisphere of the brain.

Interhemispheric Integration - Interhemispheric Integration is necessary for all higher and complex activities, such as cognitive functions, language, visual functions, multi-tasking etc.

Turning Point:

Some Pivotal Insights into Behavior and Learning

by Judith Bluestone

“Sit still and listen.”

One common mistake teachers make is to direct children to “sit still and listen.” Without getting into the technicalities of nervous system structure and function, let me explain briefly why, for some children, this statement is contradictory. There are actually several explanations, depending on the nature of the movement the child is exhibiting.

Scenario 1

The behaviors you see: The child is stretching, yawning, pushing off from the table, bouncing her head around, rocking from side to side in her chair, getting up and jumping from time to time, or engaging in other fast and sudden movements. She is telling you something important.

What this might mean: This child is saying that her vestibular functions¹ are too weak to simultaneously serve awareness of body in space, looking, and listening. She knows that what you really want and what is important for her to learn is that she listen. And so she moves and bounces to keep her vestibular system alert, energized, so she can listen. If she were to heed the first part of the direction and sit still, then she would tune out, and miss the learning that is taking place.

What you can do: If the movements are truly bothersome to the class (not just to the teacher), then build in some form of movement before and during any activity that requires sustained listening. The children might jump to their seats from the doorway or from work at various centers in the room. Rhythmic body response might be built into the lesson, or you might simply request from the child to nod or shake her head vigorously every time she agrees or disagrees with a statement. You should also probably refer the child for sensory-integration therapy or to a program that will provide her activities that will strengthen her weak vestibular functions.

Scenario 2

The behaviors you see: The child just can't sit still. He is always squirming in his chair, and more frequently than not has only the slightest bodily contact with the chair. Occasionally he appears to sit on the chair, and then immediately puts one leg under him. He then periodically shifts the leg underneath him, as it begins to fall asleep from his body pressure. He probably also keeps playing with his clothes. All his teachers complain that he can't sit still. Yet if he stretches out to listen to a story or watch a program on television, he is quiet.

What this might mean: This child probably can't sit still because he is hypersensitive to touch, particularly in the area along his sciatic nerve (buttocks and legs). The fabric of his clothing rubbing against the chair and into the back of his leg (especially behind his knee) is downright ticklish. He may not even realize this, since he has been trying to block that sensation and pay attention to the lesson for most of his life. He might be able to sit a little stiller on some days--maybe he is wearing softer or tighter clothing, maybe his diet has not aggravated an allergic skin response, etc.

What you can do: If the child has no serious problems of visual or auditory acuity, "seat" him in the back of the room, and allow him to stand, or to move in his seat as needed. That will minimize the distraction of his activities for the rest of the class. Suggest that his family get help to desensitize him through diet control, comfortable clothing, special massage, etc.

Scenario 3

The behaviors you see: The child is sitting relatively still, but her hands are not. They are frequently drumming on the table, clicking retractable pens, in her mouth, pulling on her face near her mouth. It is disturbing to those around her, and her books and papers are always a mess since her hands move directly from inside her mouth onto the papers.

What this might mean: This child is telling us that she is being sensorily overloaded by the listening activity. She wants to sustain listening so that she will learn, and so that she will be part of the group. Her body is seeking a means to balance the sensory over-load. It is providing her intense sensory-cortex stimulation through the highly represented areas of fingertips and mouth. This is counter-balancing the intense auditory stimulation, and allowing her to stay with the lesson.

What you can do: Allow the child to use an agreeable fidget toy, such as digging holes with her fingers in an eraser, using silly putty on the surface of her desk, engaging two paper clips in various ways. Also allow her to chew something (a small wad of cloth might do if gum is disallowed). Try to punctuate the discussion with brief periods of writing or drawing so that her system can unwind. Discuss the phenomena with her parents. There are a number of new therapies that help vestibular functions not get in to overload, and some that deal specifically with concerns of auditory overload.

Scenario 4

The behaviors you see: Another child can't sit still for long. Sometime into the lesson he begins to rock back and forth in his chair. Sometimes he is rocking hard enough that his chair makes noise. You even fear that he may fall over backwards in the rocking. He seems to be tuned out while he is rocking and for a short period afterwards.

What this might mean: This child is exhibiting another reaction to auditory overload. His systems are not satisfied with finding a sensory balance through tactility, as the previous child. Or perhaps, he views those behaviors as bad. In any case, after listening for a while, he becomes overloaded. He rocks back and forth, since that is the only movement that is calming to the vestibular system. He may in fact rock enough that he tunes out, both from his listening and from his attention to body in space. If he falls over, that is a certain indication that he has

lulled his vestibular system “to sleep.” He might be able to sit still and listen a little longer on some days -- maybe his diet has not aggravated his systems, maybe he has had more sleep, or maybe he has had more quiet in other classes prior to yours.

What you can do: If the child has no serious problems of visual or auditory acuity, seat him in the back of the room, and allow him to rock in his seat as needed, being sure that the wall behind him keeps him from falling over. Understand this child’s needs, and allow him to tune out on occasion, without embarrassing him by calling his name to answer a question, etc. Try to present a synopsis of the lesson at the beginning of extended listening, so if he tunes out, he will be missing embellishment and not main points. This child also would probably benefit from treatment, similar to the child above.

Scenario 5

The behaviors you see: The child just can’t sit upright for long. She almost always starts to lean back in her chair, and stay there, balanced precariously until her chair clunks back down forward. She keeps leaning back in it, sometimes even inserting the toes of her shoes under the front of the chair to help balance it.

What this might mean: This child is telling us that her muscle tone is particularly weak. She does not have a strong enough vestibular system to support muscle tone and body-in-space, and at the same time look and listen. She decides that if she is to be a learning member of the group, listening and looking are the priorities. Therefore she conserves her energy by tipping the chair in order to support her large muscles almost effortlessly (the “lazy boy”).

What you can do: Allow the child to sit as she needs. In fact, you might even have tips put on the front legs of her chair so it leans back securely without her needing to try to balance it. If the child has no serious problems of visual or auditory acuity, “seat” her in the back of the room, if her sitting posture is truly distracting to others. Encourage the family to check on nutritional reasons why muscle tone may be weak. Also encourage them to consider sensory-integration or similar therapies to work on muscle tone through vestibular strengthening and special massage.

“Turn around and look at me when I’m talking to you.”

Another frequent complaint of parents and teachers is that children seem to purposefully look away from them when they are talking with them. Some may view this as bashful behavior, while others may feel the child is being non-compliant. Actually, there are several different neurodevelopmental reasons why a person might have to counter this request.

Scenario 1

The behaviors you see: The child is seemingly facing you, but does not seem focused on you and even appears to be thinking about something else. You tell him to “Turn around, and look at me when I’m talking to you” and he turns away, looking at something at right angles to you.

What this might mean: This child is saying that his visual system works differently than most. He is probably reliant on peripheral vision. When he is facing you, he is in fact visually tuned in to things on his sides. When he turns, and seems to look away from you, he is actually then taking you in, or viewing what you want him to look at.

What you can do: This child needs to be allowed to reposition himself so that he sees things from his peripheral vision. This may mean seating him at the end of a table, so that his turning from side to side does not disturb others seated next to him. The parents should be consulted. This child may benefit greatly from vision therapy. He might even learn to use focal vision better through guided ball sports and/or playing a musical instrument.

Scenario 2

The behaviors you see: The child faces you momentarily, and then glances away or closes his eyes. He may put his head down, or tip his head up and focus somewhere else. (The same child might frequently rub his eyes, or look away from most visual material.)

What this might mean: This child, too, is saying that his visual system works differently than most. He is probably unable to sustain convergence (focus of both eyes simultaneously) on a given target. It is very disconcerting to see faces double, or to see them disfigured and dismembered in the event that the child engages in alternating suppression of one eye or the other. And so the child looks away into oblivion. Looking at you while you speak would be distracting, and might even cause the child to giggle as he receives weird visual images.

What you can do: This child needs to be allowed to look away, after getting a quick visual take on the important visual items related to the task or discussion. He may frequently turn back to see if anything has changed, but again, cannot be expected to stay focused. If you are trying to teach this child reading, a sight vocabulary on cards flashed at a relatively high speed would help this child. This child is a logical candidate for vision therapy. Since the problem is one of visual function, and not structure or disease of the eye (for which an ophthalmologist would be the correct referral), you would not be remiss making a referral to a developmental optometrist. There are several trained developmental/behavioral optometrists in most metropolitan areas. One activity that you can “prescribe” which will increase the child’s ability to converge and sustain convergence is to have him drink through a crazy straw (several times a day). Two of our twelve pairs of cranial nerves help to strengthen vision as a result of strong sucking (or blowing) with an instrument centered between our lips. It honestly does help, and it certainly can’t hurt! Playing a wind instrument might also increase binocular convergence.

Scenario 3

The behaviors you see: The child faces you on occasion, and occasionally looks away, or covers her eyes, or puts a piece of paper between her eyes and your face (or the item you want her to attend to). She frequently seems to use her hand as a visor, and sometimes has difficulty copying or reading from books and whiteboards. She becomes fidgety during periods when the A-V equipment is being used in a darkened room.

What this might mean: This child, as well, is saying that her visual system works in an unusual manner. She is most likely sensitive to light. When you are standing against a light or bright surface, or wearing light or bright clothing, or when there are extreme contrasts of light and dark in the room, this child has visual hallucinations--things may move, disappear, appear covered with swirling colored shapes, etc. Frequently people who are sensitive to light, also have an underlying problem of binocular functions, so that each eye is processing light differently. Some people have what Helen Irlen has called Specific Scotopic Sensitivity Syndrome. Some children have both. In any case, the child is doing what she can to make sense of her visual world. She cannot sustain eye contact with such distortions, since they disturb her concentration and perception.

What you can do: This child needs to be allowed to do whatever is comfortable and acceptable to the group so she can sustain total attention. If wearing a cap with a visor is not allowed, then tipping her head, or using her hands as visors, or holding up "screens" should be accepted. The teacher or parent should try to stand away from glaring surfaces, and should wear clothes that are not terribly bright or reflective. Many synthetic materials catch the light in unusual ways, as do some patterns, such as herring-bones. Colors in the yellow family (in particular) are difficult for light sensitive persons to see when they are on a whiteboard. This child also should be referred to a developmental/behavioral optometrist for screening. She may need separate or coordinated evaluation and recommendations by a professional trained in scotopic sensitivity screening. Meanwhile, she might be aided by having her worksheets photocopied onto a pastel colored paper (usually green, blue, or gray) and using colored writing paper. An interim "therapy" of choice is again a crazy straw. Drinking through it several times a day not only strengthens convergence, it also works in a remarkable way to help the pupils of the eyes dilate and contract more responsively to light.²

Scenario 4

Behaviors you see: The child sits in a group circle, sitting straight and not bothering his neighbors until the teacher begins to hold up placards and/or begin to speak. Every time the teacher begins to make a presentation, he turns around and puts his feet on the child in front of him, and leans back onto the child behind him. At times during the circle discussion, he sits straight, sometimes reverses this posture, placing his feet on the other neighbor, but consistently adopts this posture when the teacher is talking. Of course, the teacher stops the lesson to discipline him, but to no avail.

What this might mean: This child has some reason for having a particular side turned toward the source of attention. He may have reduced visual or auditory acuity in the eye or ear on that side. Most likely you would have noticed that the child asks for repetitions of spoken communication if the problem were auditory, or has difficulty playing ball and similar feats, if the problem were visual in nature. Assuming this is not the problem, the child may simply have a severe problem of midline crossing, and he needs to have his dominant side facing "the action."

What you can do: Of course, you should consult the child's health records and the child's parents to rule out sensory loss on the suspected side. Try seating the child in such a way that his opposite side is toward "the action." In addition, you should consult with the parents, and consider occupational/physical therapy or other structured activities that would

encourage midline crossing. During activities that are not important to academic achievement, try to encourage the child to rotate through and reach across the midline. But don't forget to provide compensation during activities where academic learning and self-esteem issues are involved.

What These Scenarios Tell Us

In all of these scenarios, the children involved are not being obstinate. They perceive your direction as setting them up for failure, and try to salvage what they can. They rightly perceive that their main task is to focus on the auditory or visual demands in whatever way they can, so they can learn. Their behaviors only help show us what irregularities exist in their processing. With that understanding, we can be more flexible in our approach to these children. They will respond enthusiastically. They have not been understood up to this point. And they have not understood that other people do not perceive things as they do, but just tolerate them better. Some are willing to adopt our perceptions of their behavior as being naughty, although they know that it was not intentional. No, these children are not non-complaint, nor do they have a low frustration tolerance. They experience extreme daily frustrations, and when they try to compensate themselves for them, they are berated as behaving improperly. With understanding, we can turn this situation into a win-win!

One General Rule

Rhythm and music not only stimulate the vestibular system, they also enhance memory. Try to incorporate them whenever possible.

Footnotes

¹ **Vestibular functions** refer to activities related to and dependent on reception of and adaptation to stimulation of organs primarily in a labyrinth-like system of semi-circular canals, and in the otolith organs, in the inner ear. The combined perceptions of this system give us three-dimensional organization in space. Stimulation of the system is induced not only by sound, but more significantly by movement (in particular rotational movement), responses to gravity, and altered kinesthetic states. The three major functions of the vestibular system include postural control (frequently over-simplified as balance), eye movements, and conscious awareness of space. The eighth cranial nerve (vestibular) transmits the stimuli directly to the cerebellum, which is therefore integrally included in any reference to the vestibular system. Through its effect on the reticular activating system, the vestibular system also affects mood, activity level, and general focusing of attention. Of course, since the vestibular system includes the inner ear, and is stimulated by sound, listening and auditory discrimination are also included in the functions this system supports.

² The **crazy straw** therapies, and other such approaches, are the types of therapies that HANDLE recommends most frequently. They are low cost, foolproof in implementation, usually do not result in power struggles, and they actually work! One person familiar with HANDLE techniques says that what we "prescribe" most frequently is NOTEKITAL (No tech at all).



The HANDLE[®] Institute

in union with The HANDLE Institute International, LLC

Enhancing human capability through a **H**olistic Approach to **N**euro**D**evelopment and **L**earning **E**fficiency
1300 Dexter Avenue North, Suite 110 • The Casey Family Building • Seattle, WA 98109
phone (206) 204-6000 • fax (206) 860-3505 • e-mail: support@handle.org • www.handle.org

Sit Still and Listen

Additional Scenarios with Special Relevance to Autism Spectrum Disorders

Scenario A1:

The behaviors you see: The child constantly moves, generally in a rapid rhythmic rocking motion, sometimes spinning. Frequently while rocking or spinning, his hands are also flapping, usually from his wrists with his elbows locked. Every attempt to have the child sit still and listen is met with screaming and sometimes kicking and biting, too.

What this might mean: This child is demonstrating an essential conflict between wanting to shut down his inner ear system so it will not be able to take in the constant barrage of sounds and input from bodily movement. Rapid spinning and rocking forth-and-forth can accomplish that. However, the child also wants to know where his body is in space, and so he flaps his wrists and locks his elbows to fan the subtle “fire” or energy in the inner ear so he will be able to feel himself through his joints and know that he IS and that he is safe. If forced to stop these forms of self-stimulation, he may lose his sense of self and be totally overwhelmed by the information the inner ear is picking up on the sounds in his environment – input that is not under his control and probably unable to be anticipated. This could create a sense of severe anxiety.

What can you do: Reduce the intensity of sound stimulation if possible. Allow the child to engage in the self-stimulatory behaviors since they are serving an important purpose. Begin to strengthen the child’s vestibular system, gently enhancing each axis separately in a situation of security to engender trust. Explore the possibilities of allergies and intolerances to foods and environmental stimuli that may be causing a build up of fluids in the ears that could be partially responsible for the presenting behaviors. Assess other systems whose irregular functions may be interfering with the ability of the inner ear to maturely accept its full range of responsibilities to directly or indirectly regulate everything we do.

Scenario A2:

The Behaviors you see: The child is sitting, but engaging in self-stimulatory behaviors that freak out those around him and are detrimental to his own health. He continuously picks scabs from any place on his body where he had mosquito bites recently – bites which he had scratched until they bled. Any attempt to stop this behavior (with gloves or other hand restraints) causes the child to exhibit whole body writhing and sometimes kicking.

What this might mean: This child is most likely hypersensitive to touch, especially to light and ambiguous touch. The sensation of itching is an extremely annoying to anyone, particularly to those whose sensory systems seek constancy. The brain has an easier time ignoring chronic pain than it does ignoring itching sensations. Instinctively the child scratches the scabs since in the process of healing, there is movement of the various layers of skin – slightly contracted around the tighter tissue of the scab in comparison with the pliability of the normal skin tissue. This disconnected sense is intolerable. Similarly at the beginning of this problem, the child may have been guided by an

instinctive sense to protect himself from toxins in the insect bite as well as to eradicate the itch caused by the bite itself.

What can you do: This child needs to become more comfortable with light touch and with unexpected tactile experiences. While there are many forms of therapy that address this issue, it is important in choosing the right approach to remember the following basic rule: although immediately the goal may be to inhibit the receptors that are causing the over-reaction to this form of stimulation, it is ill-advised to shut down the reception of information from any of our systems (in this instance, the tactile system), since the brain needs input from all of our senses in order to develop perceptions based on the total experience. Meanwhile, there are a few measures that may help reduce the self-injurious behaviors:

- apply an unscented oil (preferably organic) and/or Vitamin E to the scabs, can soften the scab tissue so it will feel less discrepant from the rest of the skin
- provide the child with pleasant and predictable tactile experiences to keep his fingers and/or mouth busy and help the sensory-motor cortex to override unpleasant information on less highly represented body areas (arms, back, etc.) with experiences it will seek to stimulate the more highly represented areas (face, tongue, finger tips, hands).

Scenario A3

The behaviors you see: The child repeatedly taps or flicks her hand onto hard surfaces (the wall, the window, her own head). Sometimes the tapping gets almost ferocious in intensity, and other times it seems to stop without warning. Any attempt to disrupt this activity is met with a forceful continuation of the movement until it seems to stop on its own.

What might this mean: The child, like a doorbell, has had his buttons pushed (although the child's figurative buttons can be anything in his internal make-up that is jangled by certain sensory input). Once the button has been pushed, neural energy surges through the system seeking release, and like the doorbell, once contact has been made, the transmission of the signal has achieved its end and stops. However, if the button is again pushed, again and again, then the child needs to continue to make sharp and firm contact in order to stop the movement of aberrant neural energy through his system. So, this again is simply a sign that the child's systems are stressed.

What can you do: Determine what it is that is "pushing the buttons" and work to integrate the senses so they can accept the sensory input without it causing aberrant energy. As a temporary compensatory measure, try to protect the child from the offending stimuli. It may also help to reduce the amount of synthetics in the clothing and toys and total environment, since synthetics do not allow for a release of energy. Also, for some individuals, having copper based ointments on the soles of their feet or copper ankle bracelets on their ankles may help, since copper is "the grounding wire." For children who thrash a lot in their sleep, it sometimes helps to wind a copper wire from the bed frame down the foot of the bed to the floor to help discharge aberrant energy.



Section 31

Breakout Session #8
Friday, July 27, 2007, 3:45-4:45
Pacific Ballroom IV

A Wholistic Approach to
Developing Education
Plans



Presented by
Donna Lynne
and Kim Zeigler

8th International CHARGE Syndrome Conference, Costa Mesa, CA, July 26-29, 2007

A Wholistic Approach to Developing Education Plans

Donna Lynne, Technical Assistance Coordinator
Kim Zeigler, Family Services Coordinator

University of Kentucky
Department of Special Education and Rehabilitation Counseling

Kentucky Deaf-Blind Project

July 28, 2007

Presentation Objectives

- To provide participants with the tools they need in order to pull together an effective team that can:
 1. Identify specific medical and health issues that impact the child's learning;
 2. Identify specific OT, PT, SLP, Vision and Hearing issues that impact the child's learning;
 3. Identify specific communication needs that impact the child's learning;
 4. Identify what works and what doesn't work, thereby
 5. Developing a *wholistic* education plan that addresses a child's unique learning needs

IFSP/IEP Meeting ... Realities

- Time is often limited ... So completing paperwork becomes the number one objective
- People in attendance give their separate reports ... but little attention is given to viewing the child from a *wholistic* perspective

IFSP/IEP Meeting ... Realities

- Assistants, paraprofessionals, other family members, students/friends are usually not at the meeting and often receive information from the meeting second hand or in fragments
- Each individual leaves with his/her designated goals/objectives (assignments)

Realities (continued)

- Goals/objectives are written in such a way that they are carried out in isolation, out of context and easily become non-functional
- The family leaves with a completed document that contains a great deal of information that may or may not include:
 - WHAT the child will learn
 - WHERE the child will learn and
 - HOW the child will learn

Challenges To Meet Because

- Teachers are very, very busy ... hard to find time to meet with them
- The paraprofessionals are the ones who have the most contact with the students
- Misunderstandings and fragmented information impedes the best intentions
- People working with the student *do* want to do their best and be a positive influence ... but, often they don't know how

What Team Members NEED to Know

- The child's unique learning characteristics specific to the seven sensory systems
- Strategies for calming or arousing the child in order to bring him/her to the optimal state for learning and engagement
 - How to read the child's regulatory state "Ready, Set, Go" for learning

What Team Members NEED to Know

- The child's Expressive communication including both form and function
- The child's Receptive communication including both form and function

What Team Members NEED to Know

- How to gain the child's attention (which should precede all other receptive functions)
- How to greet the child
- How to let the child know that you are leaving

What Team Members NEED to Know

- The daily schedule of routines and activities for home and school (the context for instruction)
- Task analysis of each routine/activity that reflects all four (4) parts of the routine/activity
 - Initiation, Preparation, CORE, Termination*
- If the schedule needs to be modified it should include a balance between high energy and low energy activities

What Team Members NEED to Know

- The individualized calendar or schedule system the child requires
- Protocol for implementing the system
- Most current and updated information on vision and hearing
- Toileting routines
- Characteristics of CHARGE Syndrome as they are manifested in the individual child

Determine which pieces of information need to be included as part of Specially Designed Instruction !!!

Strategies for Sharing

○ Videotaping

- Document progress over time across environments
- Share between home and school
- Share during formal meetings to elaborate on key points
- To educate/inform new personnel
- To share with family members

Strategies (continued)

○ Customized Communication Form

- To share information between all team members, including home and school
- Addresses specific information based on family priorities and concerns
- Utilizes a format agreed upon by all team members

Strategies (continued)

○ Customized Communication Form

- Facilitates ongoing communication
- Disseminated using a variety of formats, including e-mail

Strategies (continued) ...

○ Electronic Communication

- Distribution list of all team members
- Keeps everyone in the loop
- Facilitates sharing of a variety of information
- Ability to share info including daily communication form

Strategies (continued) ...

○ Team Meetings

- Needs to be agreed upon and documented in the IFSP/IEP Conference Summary
- Needs to be planned for in advance, regular and consistent versus reactionary
- Must be purpose driven (agenda disseminated prior to meeting)

Strategies (continued) ...

○ Team Meetings

- Determine who needs to be included
- Assigned roles (ex: facilitator, recorder)
- Request PD hours for staff

Strategies (continued) ...

Team Meetings

- **Development of Action Plan at end of each meeting**
- **The Action Plan incorporates collaboration strategies for participation of all team members**
- **Agree upon date for next meeting before leaving**

Contact Information

- o Donna Lynne
229 Taylor Education Building
Lexington, KY 40506-0001
Donna.Lynne@nkces.org
859-322-1301
- o Kim Zeigler
229 Taylor Education Building
Lexington, KY 40506-0001
kzeigler1@msn.com
502-418-8189