

THE AIRWAY IN CHARGE: FOR THE PHYSICIAN

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Airway management is often one of the major problems in CHARGE. Many of the anomalies are well-known entities with well-established management protocols. However, CHARGE has several fairly unique features which make decision-making difficult, particularly whether or not to do a tracheotomy.^{1,2}

ANOMALIES

Choanal atresia – surgical management is not different from other conditions.³

Laryngo-tracheo-bronchomalacia – can be severe⁴ leading to collapse of bronchi and decreased air movement.

Cleft lip and palate – surgical management is standard but feeding management is not. See gastroesophageal reflux below.

Gastroesophageal reflux related to neonatal brain stem dysfunction, which is the major cause of feeding problems in CHARGE. NBSC, previously reported in children with Pierre Robin sequence is a group of four types of symptoms involving the supranuclear region of the IXth, Xth and XIIth cranial nerves.^{5,6,7} These symptoms, important in the first two years of life, seem to be related to a developmental defect of the suck-swallow central pattern generator in the solitary tract.⁸ Recurrent aspiration pneumonia due to this problem is a major cause of morbidity and mortality.⁹

Heart disease – anomalous pulmonary venous return and vascular rings have been reported.¹⁰

Tracheoesophageal fistula – surgical management is the same but, again, feeding is not.

Other more minor airway anomalies and those above which are not recognized prior to anesthesia can lead to difficulty with intubation.¹¹

Recent anecdotal report of asymmetry of diaphragm movement was noted on fluoroscopy of a one-year old.

MANAGEMENT DECISIONS

Tracheotomy: While most physicians try to avoid tracheotomy in these patients, the swallowing problem can be so severe that the child does not swallow its own secretions. The secretions, then, become copious and are a management problem in themselves. They obstruct the upper airway and may be aspirated along with any feedings. After tracheotomy

is performed, secretions and any formula or food is usually seen coming out the trach with suctioning. Tracheotomy may be performed in a child who has NBSC until such time as it resolves spontaneously usually over several years. Rarely is there a need for permanent tracheotomy, but several cases are known.

Tracheal diversion: This procedure has been performed in older children whose NBSC has not resolved spontaneously, are unlikely to develop speech, and who show a strong desire to eat.

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