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Management of upper airway obstruction in Pierre Robin sequence

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Abstract
Pierre Robin sequence (PRS) presents in the neonatal period with upper airway obstruction and feeding difficulties. Infants with pronounced micrognathia may fail to thrive because of chronic airway obstruction, or experience severe respiratory distress. This is potentially fatal and surgical intervention in these cases is necessary. We present our series of cases with severe PRS requiring surgical relief of their airway obstruction, and the reasons for preferring tracheostomy over glossopexy.

Key words: Airway obstruction; Pierre Robin syndrome

Introduction

The diagnostic criteria of PRS – micrognathia, glossoptosis, and respiratory obstruction usually associated with an incomplete cleft palate – are well known (Robin, 1934). It can present as an isolated anomaly or may represent part of a syndrome (Lewis and Pasyayan, 1980; Pasyayan and Lewis, 1984; Shprintzen, 1992). The incidence of PRS is reported to be 1:8,500 (Bush and Williams, 1983).

There is a wide variation in the severity of PRS which presents in the neonatal period with upper airway obstruction and feeding difficulties. These symptoms gradually subside as mandibular growth and improved control of the tongue musculature develop (Pruzansky and Richmond, 1954).

Airway obstruction in mild cases can be managed conservatively by nursing the infant in the prone position. In more severely affected infants, a nasopharyngeal prong can be used to bypass the pharyngeal obstruction caused by glossoptosis. This technique is useful for short periods of time but is not suitable for lengthy periods of airway support as the tip of the prong can cause irritation or vomiting, or become displaced into the oesophagus. Feeding difficulties may be overcome by fashioning orthodontic teats, insertion of a nasogastric tube, or even gastrostomy.

However, in neonates with pronounced micrognathia, failure to thrive due to chronic airway obstruction, or severe respiratory distress may ensue, despite the above measures. This situation is potentially fatal (Robin, 1934; Douglas, 1950). Surgical intervention to relieve the obstruction in these cases is necessary. This may take the form of a glossopexy or tracheostomy.

We present our series of cases with severe PRS requiring surgical intervention and the reasons for preferring tracheostomy over glossopexy.

Materials and methods

The records were reviewed of all infants with PRS requiring tracheostomies at the Sheffield Children's Hospital from 1982 to 1997.

All patients were initially nursed on a 'Special care baby unit' with intensive care facilities. The management of these infants included nursing in the prone position, nasopharyngeal prongs and endotracheal intubation for airway support. Feeding difficulties were managed with orthodontic teats, nasogastric tubes and gastrostomy.

Only if infants failed these supportive measures or had life threatening airway obstruction was a tracheostomy performed. In all cases, these infants were assessed, and had the tracheostomy performed, by a consultant otolaryngologist with a special interest in paediatric airways.

Results

Seven infants with PRS over this 15-year time period required tracheostomy. There were four males and three females. Two were premature births. One infant had deletion of the long arm of chromosome 4, and another had Larsen's syndrome. Two infants could feed normally, four required nasogastric tube feeding and one had a gastrostomy (Table I).

In all patients intubation was extremely difficult. One patient was impossible to intubate and the anaesthetic was delivered through a rigid Storz

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TABLE I
PIERRE ROBIN SEQUENCE TRACHEOSTOMIES

Patient	Gestation	Syndrome	Feeding	Age at tracheostomy	Age at cleft palate repair	Age at decannulation
Male	31		Gastrostomy	126 days	1 yr 8 mths	1 yr 9 mths
Male	33		NG tube	46 days	5 mths	1 yr 1 mth
Male	40	Deleted chrom. 4	Normal	10 days	1 yr	3 yrs 5 mths
Female	40		NG tube	27 days	1 yr 4 mths	7 mths (fell out)
Male	39		NG tube	114 days	1 yr 2 mths (soft cleft) 1 yr 6 mths (hard cleft)	3 yrs 1 mth
Female	40		NG tube	22 days	10 mths	2 yrs 2 mths
Female	41	Larsen's	NG tube	32 days	1 yr 9 mths	2 yrs 7 mths

bronchoscope. The age at which tracheostomy was performed ranged from 10 days to 126 days.

In all patients the tracheostomy was left *in situ* until after they had had definitive repair of their cleft palate, except in one case in which accidental decannulation at home occurred.

The age at decannulation ranged from seven months (accidental decannulation) to three years five months. Five infants had to undergo closure of a tracheocutaneous fistula. There have been no mortalities.

Discussion

In 1946, Douglas, reporting on the findings of laryngoscopic examination in infants with PRS, thought that micrognathia causes the tongue, which is relatively unsupported due to the diminished axis of the genioglossus, to fall backwards acting like a ball valve on inspiration to obstruct the hypopharynx (Douglas, 1946). This situation is worsened by the effect of gravity on the tongue when the infant is supine, and also when the tongue becomes impacted in the cleft palate (Routledge, 1960).

In his original article, Douglas described a glossopexy procedure, where the tongue was effectively 'anchored' forwards. In a subsequent review of 25 cases of PRS treated with this procedure, he reported a 100 per cent survival rate (Douglas, 1950). This compared favourably to a 35.7 per cent survival rate in cases that were managed conservatively, and showed the benefit of surgical intervention in these children.

As regards tracheostomy, this was not thought to be a logical surgical option in the management of these children owing to practical concerns regarding the management of paediatric tracheostomies of this era. In particular, all young children were hospitalized until the tracheostomy was removed, and a significant mortality was associated with this situation (Hawkins and Williams, 1976; Line *et al.*, 1986). This compared to infants who went home within a few weeks having had an uncomplicated glossopexy. In view of these results, glossopexy became generally accepted as the procedure of choice for infants with severe PRS.

However, the popularity of this procedure has declined. Reports that glossopexy could not always provide a resolution of symptoms in PRS appeared in the literature (Routledge, 1960; Augarten *et al.*, 1990). It also became apparent that a number of complications such as tongue lacerations, wound infections, dehiscences, injuries to the submandibular ducts, and scar deformations of the lip, chin and floor of mouth, often required revision procedures for correction (Routledge, 1960).

More recently, it has been recognized that, in a number of cases, PRS represents part of a syndrome in which there is an associated neuromuscular or developmental disorder which may not be diagnosed until later in life (Lewis and Pasyayan, 1980; Pasyayan and Lewis, 1984; Shprintzen, 1992). The pathophysiology of upper airway obstruction has been further clarified by Sher *et al.* who performed flexible fibreoptic endoscopy in these infants (Sher *et al.*, 1986). Four distinct types of upper airway obstruction have been described:

Type 1 The obstruction consists of the posterior movement of the dorsum of the tongue to the posterior pharyngeal wall so that the majority of airway obstruction is anteroposterior.

Type 2 The tongue moves posteriorly, but instead of contacting the posterior pharyngeal wall, the tongue compresses the soft palate or the cleft palate tags posteriorly against the posterior pharyngeal wall so that there is a junction of the tongue, velum and posterior pharyngeal wall in the upper portion of the oropharynx.

Type 3 The lateral pharyngeal walls move medially causing them to appose one another.

Type 4 The pharynx constricts in a circular or sphincteric manner with movements occurring in all directions.

In a series of 53 cases of PRS evaluated using this technique to determine the most appropriate surgical intervention, it was found that Type 1 obstruction (58.5 per cent) responded well to glossopexy (Argamaso, 1992). However, in Types 2, 3 and 4 (39.6 per cent) tracheostomy was indicated (Sher, 1992).

Since glossopexy was originally described, mortality figures associated with paediatric tracheostomies have reduced dramatically and long-term management of these cases is now at home as opposed to hospital (Carter and Benjamin, 1983; Crysdale *et al.*, 1988; Freezer *et al.*, 1990; Shinkwin and Gibbin, 1996). It has also become recognized that if other techniques have failed, tracheostomy will successfully bypass the obstruction in PRS (Augarten *et al.*, 1990).

In most ENT Departments, the number of infants with PRS requiring surgical intervention is small. In our own unit which has a special interest in paediatric airways, only seven cases with PRS

requiring tracheostomy were seen over a 15-year period. Therefore, experience in assessing airway obstruction in an infant with PRS using flexible fibreoptic endoscopy, to decide whether glossopexy or tracheostomy is more appropriate, would be extremely limited. These children have precarious airways and even attempted intubation is a potentially fatal event. In view of the small number of cases involved, we consider the safest option is to perform a tracheostomy. Not only does this guarantee that the upper airway obstruction is bypassed, but it also allows future general anaesthesia, necessary for surgery on the palate, to be given through the tracheostomy.

Conclusion

Infants with PRS who have upper airway obstruction not responding to conservative measures should be assessed by an otolaryngologist with a special interest in paediatric airways. Only if experience has been obtained of assessing the mechanism of upper airway obstruction using flexible fibreoptic endoscopy in these children should consideration be given to glossopexy. We advocate that unless this experience is available the safest option to relieve the upper airway obstruction is tracheostomy. This also offers access to the airway for future general anaesthesia when necessary for palatal surgery.

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