Oesophageal Atresia Treatment: A 21st-century Perspective

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The importance of the following 3 general aphorisms needs to be considered:

1. There is a well-defined and clear relationship between volume (of cases), management outcome, and research output.
2. Most medical and surgical procedures have a better outcome when performed in hospitals that do a lot of the procedure in question.
3. Increased hospital specialisation is associated with improved patient outcomes.

A clear example of the effect of centralisation of care is the outcome for biliary atresia in the United Kingdom. Before 1995, 15 centres were performing surgery for biliary atresia. The 5-year native liver survival in the 2 centres with ''high'' volume was 61% compared with 14% in the 13 centres operating on fewer than 5 cases per year. This led the National Health Service to legislate that in the future, only 3 centres would be designated to carry out surgery on biliary atresia. The results for biliary atresia in the 3 centres since their designation as special centres continues to be highly satisfactory, with a 89% overall survival and a 5-year survival rate with native liver of 51% (1). Recently, 6 other paediatric surgical conditions have been allocated supraregional status. The common theme for all of them is volume, specialisation, and the team approach.

Oesophageal atresia is a relatively uncommon condition—1 in 2500 to 3000 births. Eighty-five percent of cases are of the common variety of oesophageal atresia with distal tracheooesophageal fistula. Each regional centre, serving a population of 2.5 million, will manage on average 8 to 10 cases per year. With 5 paediatric surgeons per centre, each surgeon will operate on 1 to 3 cases of oesophageal atresia per year. For isolated atresias, which make up only 8% of cases of oesophageal atresia, surgeons will manage only 1 case every 5 to 10 years. This is clearly unacceptable.

The management of long-gap atresia (defined as atresia without distal fistula) is complex and can be divided into 3 stages: at the initial procedure with anastomosis under tension with or without elective paralysis and mechanical ventilation, delayed primary repair or use of the Foker technique, and replacement procedures. It is important to recognise that 10% to 15% of so-called isolated atresia have a proximal fistula.

Every effort should be made to retain the patient's oesophagus, but ''persistent futile attempts to retain the native oesophagus can have disastrous effects on the child and the family'' (2).

Gastric transposition is now accepted as the replacement procedure of choice in many centres. We have carried out the procedure in 192 cases (1980–2005) with a 4.6% mortality rate (a reflection of the extremely complex nature of the cases referred to our centre nationally and from abroad), a leak rate of 12%, and stricture of 20% (mostly following caustic ingestion) (3). A highly satisfactory outcome was achieved in 90% of cases, and there was no deterioration of function over time.

Recurrent fistula occurs in 5% to 8% of cases, so any 1 centre will only manage 1 or 2 cases in 10 years. The repair of the recurrent fistula can be challenging and the rerecurrence rate is 10%.

Tracheomalacia in its mild form is common and is responsible for the ''barking cough'' characteristic in children with repaired atresia. In its severe form it causes apnoeic and/or cyanotic attacks. It is caused by defective cartilaginous support of the trachea mainly at the site of the fistula. Aortopexy is dramatically effective in resolving the problem (4).

What measures would I suggest to improve the outcome for oesophageal atresia?

1. Centralisation and concentration of patients particularly for
   a. Pure and long-gap atresia
   b. Replacement procedures
   c. Recurrent trachoeoesophageal fistula
   d. Severe tracheomalacia
   e. Associated major cardiac defects
2. Recognition of the value of teamwork and specialisation in
   a. Paediatric anaesthesia
   b. Intensive care
   c. Cardiac, renal, urological, and orthopaedic expertise
   d. Expert radiology
   e. Respiratory support
3. Careful and long-term follow-up in multidisciplinary teams with particular reference to respiratory and swallowing problems, nutritional support, and early recognition and treatment of gastrooesophageal reflux, strictures, tracheomalacia, and recurrent fistula
4. Promotion of the work of parent support groups, which provide knowledge of day-to-day problems, psychological support, and occasionally financial assistance, and raise money for research and development

REFERENCES

1. Davenport M, De Ville de Goyet J, Stringer MD, et al. Seamless management of biliary atresia in England and Wales (1999–2002). *Lancet* 2004;363:1354–7.
2. Holland AJ, Ron O, Pierro A, et al. Surgical outcomes of esophageal atresia without fistula for 24 years at a single institution. *J Pediatr Surg* 2009;44:1928–32.
3. Spitz L, Kiely E, Pierro A. Gastric transposition in children—a 21-year experience. *J Pediatr Surg* 2004;39:276–81.
4. Spitz L. Esophageal atresia—lessons I have learned in a 40 year experience. *J Pediatr Surg* 2006;41:1635–40.

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