Case Reports

Esophageal-Aortic Erosion Associated with Double Aortic Arch and Tracheomalacia

Experience with 2 Infants

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Patients with double aortic arch may require lengthy intubation for ventilatory support. The need for endotracheal and nasogastric intubation may be prolonged in such patients because of associated tracheomalacia. Iatrogenic tracheal or esophageal erosion with subsequent aortic fistulization is an unusual but catastrophic complication that may result from such intubation.

We report the cases of 2 infants with double aortic arch and tracheomalacia who developed iatrogenic esophageal-aortic erosion. This complication was successfully managed in 1 of the infants. We conclude from our experience that the important steps in preventing this complication include 1) expediting the exclusion of upper-airway compromise in intubated infants who have a presentation characteristic of bronchospastic airway disease (hyperinflation and hypercapnia) that seems unresponsive to usual therapeutic measures; and 2) expediting the diagnosis of vascular ring in order to minimize the duration of dual tracheal and esophageal intubation. Effective management of this problem, once established, requires primary closure of the esophageal perforation, removal of the nasogastric tube, interposition of thick viable tissue between the esophogus and the aorta, and decompressive gastrostomy and feeding jejunostomy. Concomitant aortopexy may be appropriate. ***(Texas Heart Institute Journal 1993;20:126-9)***

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Esophageal erosion as a complication of prolonged intubation in association with congenital vascular ring, especially in conjunction with double aortic arch, has been documented, albeit rarely, in several large series.[1-6] We report our recent experience with 2 infants with double aortic arch and tracheomalacia who developed iatrogenic esophageal-aortic erosion. The details of patient presentation and the factors that predispose to the development of this complication are emphasized in the hope that increased awareness might prevent similar occurrences in the future. We present simple and effective technical modifications to the surgical management of this problem.

Case Reports

Patient 1

A 6-week-old black male was admitted on an emergency basis to our institution on 5 December 1988, because of a prolonged apneic episode. He had been delivered by cesarean section at 8 months' gestation due to maternal placenta previa and had required brief intubation the next day for respiratory distress. He had been discharged 5 days later with persistent respiratory distress and stridor and with the diagnosis of hypoplasia of 1 lung. During subsequent weeks, he had exhibited persistent respiratory and feeding difficulties and a chronic cough.

The day after his admission to our institution, the patient required endotracheal and nasogastric intubation because of another apneic episode. During the next 3 weeks, he experienced hyperinflation, hypercapnia, and inadequate ventilation, and he was presumed to have bronchospastic airway disease. The condition responded poorly to ventilator support and pharmacologic interventions. On 28 December 1988, his endotracheal tube was replaced, at which time it was noted that adequate ventilation occurred only when the tip of the endotracheal tube was

located at the tracheal carina. Upper-airway compromise was objectively confirmed by characteristically abnormal airway pressure-volume dynamics on the ventilator. Cardiac catheterization confirmed a right-dominant double aortic arch, and immediate surgery was undertaken.

The hypoplastic anterior left arch and ligamentum were divided. Mobilization of the esophagus revealed mucosa adherent to the posterior right arch and a contained esophageal perforation. The perforation was repaired and covered with a free autogenous pericardial graft. The nasogastric tube was left in place. On the 7th postoperative day, retraction of the endotracheal tube was accompanied by inadequate ventilation, suggesting persistent distal upper-airway collapse. Six days later, he began passing bloody stools. He was taken to emergency surgery the following day, where the source of the hemorrhage was found to be an aortoesophageal fistula. Attempts at repair were unsuccessful, and the patient died of exsanguination in the operating room.

Patient 2

A white female infant, born by vaginal delivery at 34 weeks' gestation, required 36 hours of intubation thereafter for respiratory distress. Post-extubation stridor prompted predischarge bronchoscopy, and a subsequent diagnosis of laryngomalacia was made. The patient had persistent respiratory and feeding difficulties and intermittent cough. On 4 April 1992, at 3 weeks of age, she was admitted to another hospital following a prolonged apneic episode. Barium esophagograms performed there were initially interpreted as normal (Fig. 1A); however, she continued to have intermittent apneic episodes. These culminated in respiratory arrest on 28 April 1992, which required endotracheal and nasogastric intubation.

During the next 3 weeks, the patient had substantial difficulty with hyperinflation, hypercapnia, and inadequate ventilation. She responded poorly to ventilatory and pharmacologic interventions for presumed bronchospastic airway disease. Adjustment of the endotracheal tube on 18 May 1992, with subsequent juxta-carinal placement, was correlated with improved ventilation; therefore, extrinsic upper-airway compression was suspected. She was transferred to our institution, and cardiac catheterization confirmed a symmetric double aortic arch (Fig. 1B) for which she underwent emergency surgery.

Distal division of the left anterior arch and ligamentum was performed through a left thoracotomy, and mobilization of the esophagus revealed a contained posterior esophageal perforation with partial erosion of the posterior right arch (Fig. 2A). The nasogastric tube was removed, the perforation was closed, and the eroded arch was imbricated with a longitudinal adventitial suture, of necessity creating

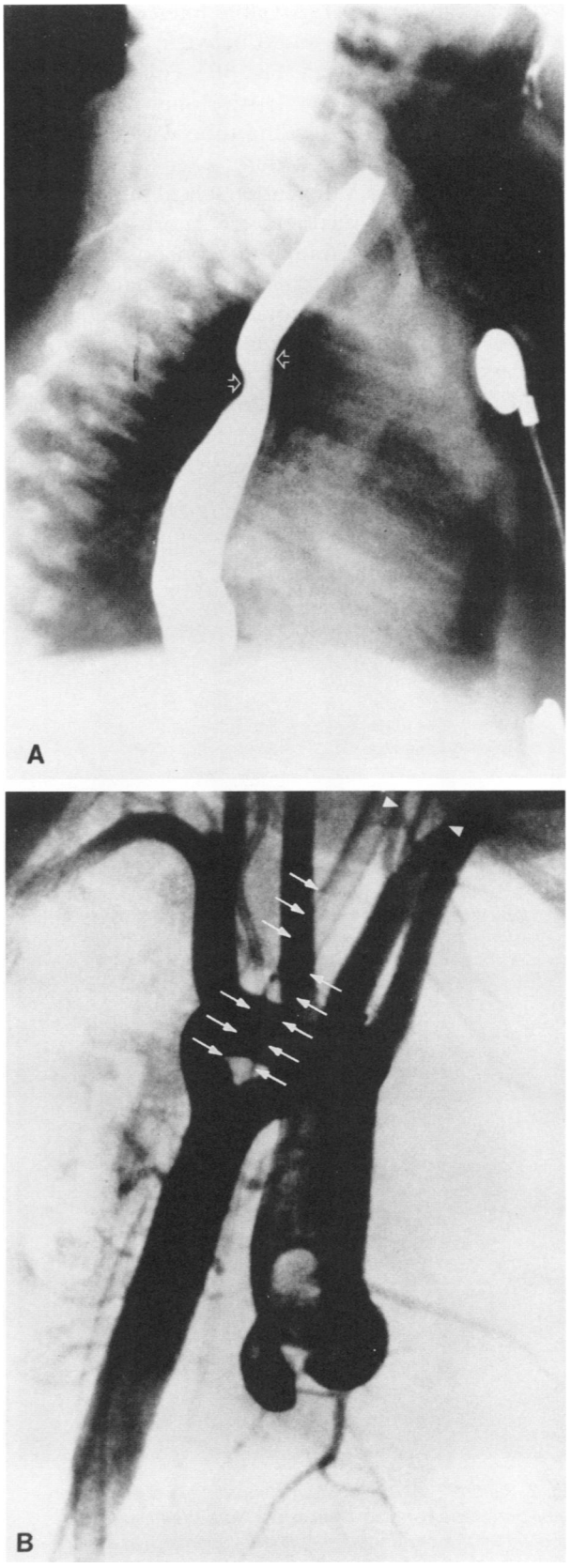

Fig. 1 *Patient 2.* ***A****) Barium esophagogram showing anterior and posterior indentations caused by double aortic arch (arrows), and* ***B****) aortogram. Note the nasogastric tube (small arrows) and the endotracheal tube (arrowheads).*

a coarctation (Fig. 3A). A viable intercostal muscle pedicle was then interposed between the repaired esophagus and the aorta (Fig. 3B). Following chest closure, a decompressive gastrostomy was accomplished and a jejunal feeding tube was placed via separate transgastric procedure.

Attempts to retract the endotracheal tube postoperatively resulted in recurrent ventilatory inadequacy and hyperinflation, prompting bronchoscopy on 9 June 1992, which confirmed tracheomalacia (Fig. 2B). Pericardial flap aortopexy was subsequently performed through a right anterior thoracotomy, and extubation was successfully accomplished several days following this 2nd procedure. An initial post-repair coarctation gradient of 65 mmHg had improved to 10 mmHg at discharge 4 weeks later.

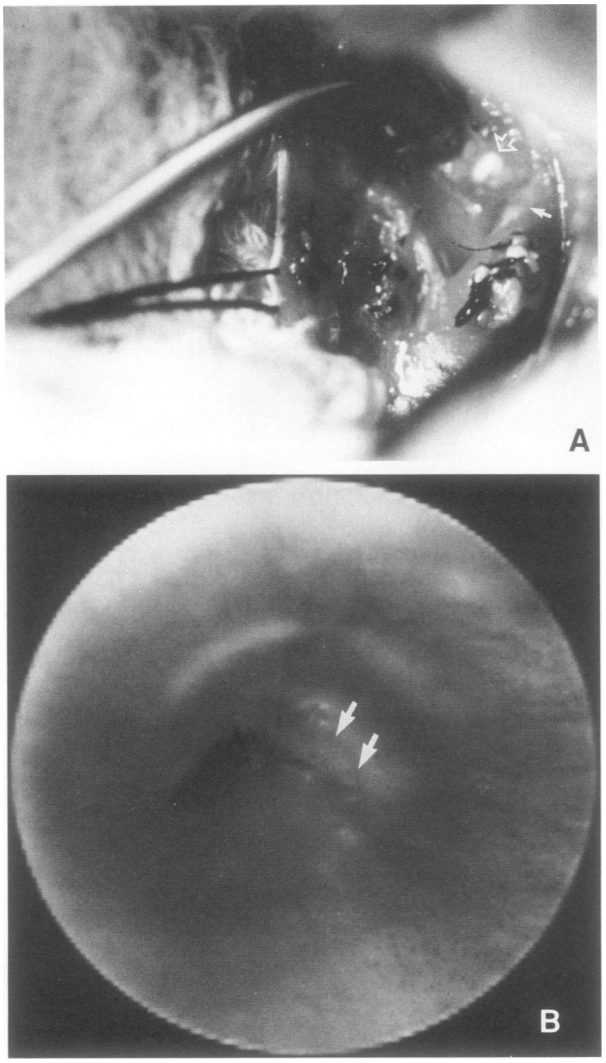

Fig. 2 *Patient 2.* ***A)*** *Intraoperative photograph of the esophageal perforation and aortic erosion. Note the nasogastric tube within the perforated esophagus (open arrow) and the "target" effect created by its erosion of progressive layers of the posterior right aortic arch wall (solid arrow).* ***B)*** *Postoperative telescopic bronchoscopy demonstrating tracheal collapse and obliteration of the right main bronchial orifice (arrows) due to tracheomalacia.*

Discussion

Mediastinal visceral erosion as a complication of double aortic arch has been previously correlated with the need for prolonged ventilatory assistance. Effective initial control of this complication, however, has not met with success. Chun and associates,[1] reviewing the Johns Hopkins experience, reported an unsuccessful attempt to repair a tracheoaortic fistula that resulted from a tracheostomy tube necessitated by an undiagnosed vascular ring. Arciniegas and colleagues[2] described a case of aortoesophageal erosion in an infant who had required ventilatory support for 6 weeks after surgery. The infant underwent primary repair of the erosion 19 days after vascular ring division, but erosion recurred 9 days later. The authors do not report whether this patient

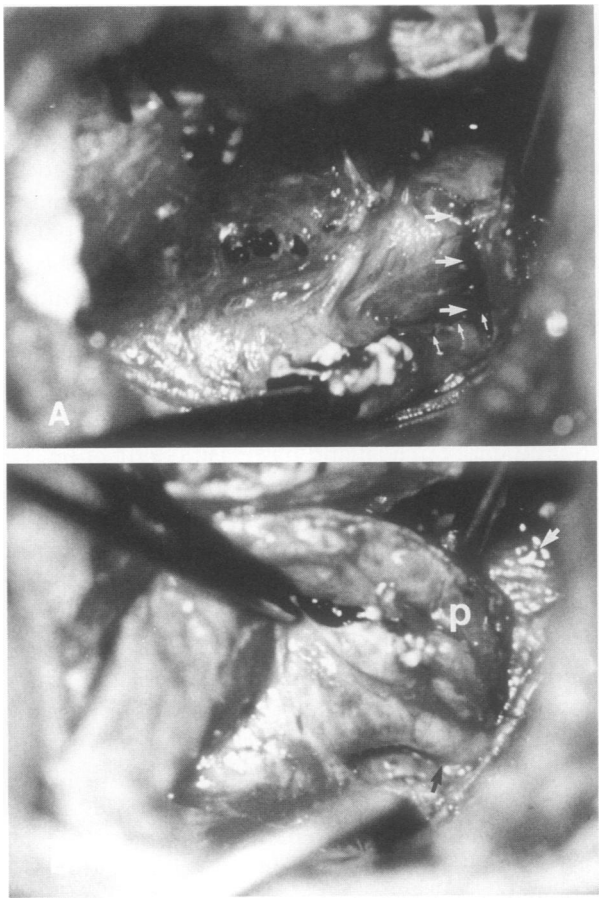

Fig. 3 *Patient 2. Operative photographs demonstrating* ***A)*** *the closed esophageal perforation (large arrows) and the imbricated posterior right aortic arch erosion (small arrows), and* ***B)*** *the intercostal muscle between the pedicle flap (p) sutured between the repaired esophagus (white arrow) and the posterior aortic arch (black arrow).*

required extended postoperative intubation, which might have contributed to this event. Successful outcome was ultimately achieved with esophageal exclusion and delayed reconstruction.

The similarity between our 2 patients emphasizes several important points. First, classic symptoms of upper-airway and gastrointestinal compromise should mandate early examination for vascular ring even though this entity represents an infrequent cause of infant respiratory distress. Second, once intubation of the trachea and esophagus has been necessitated by an apneic event, possible upper-airway compromise should be investigated promptly—especially if the intubation is associated with intractable ventilatory difficulty, hyperinflation, and hypercapnia. This investigation may be accomplished objectively with airway pressure or flow dynamics from the ventilator,[7] or subjectively by assessing ventilatory adequacy with variable positioning of the endotracheal tube relative to the tracheal carina. Should either assessment suggest upper-airway compromise, definitive diagnostic techniques such as magnetic resonance imaging or cardiac catheterization should be applied to look for a vascular ring.[8] Third, physicians should recognize that dual intubation of the mediastinal viscera within the tight confines of a double arch that is restrictive enough to cause severe symptoms (e.g., reflex apnea) predisposes the patient to early visceral erosion in a relatively short period of time due to the dynamics of pulsatile flow against a rigid stent.

Finally, when a patient has required extended preoperative intubation, mobilization of the esophagus following division of the vascular ring should be undertaken to investigate whether or not erosion of the esophagus has occurred. If such a lesion is found, the nasogastric tube should be removed and bulky viable tissue interposed between the repaired esophagus and aorta to avert delayed catastrophic hemorrhage. Providing a decompressive gastrostomy and a feeding jejunostomy facilitates control of post-repair reflux and expedites healing, and obviates continued pressure necrosis of the esophagus. These steps are particularly important because division of the vascular ring may not relieve upper-airway obstruction that is due to associated tracheomalacia. Such failure may necessitate extended postoperative intubation, secondary aortopexy, or both.[9-12]

References

1. Chun K, Colombani PM, Dudgeon DL, Haller JA Jr. Diagnosis and management of congenital vascular rings: a 22-year experience. Ann Thorac Surg 1992;53:597-603.
2. Arciniegas E, Hakimi M, Hertzler JH, Farooki ZQ, Green EW. Surgical management of congenital vascular rings. J Thorac Cardiovasc Surg 1979;77:721-7.
3. Bertolini A, Pelizza A, Panizzon G, et al. Vascular rings and slings: diagnosis and surgical treatment of 49 patients. J Cardiovasc Surg 1987;28:301-12.
4. Filston HC, Ferguson TB Jr, Oldham HN. Airway obstruction by vascular anomalies: importance of telescopic bronchoscopy. Ann Surg 1987;205:541-9.
5. Vallette RC, Arensman RM, Falterman KW, Ochsner JL. Tracheoesophageal compression syndromes related to vascular ring. South Med J 1989;82:338-41.
6. Backer CL, Ilbawi MN, Idriss FS, DeLeon SY. Vascular anomalies causing tracheoesophageal compression. Review of experience in children. J Thorac Cardiovasc Surg 1989;97:725-31.
7. Miller RD, Hyatt RE. Evaluation of obstructing lesions of the trachea and larynx by flow-volume loops. Am Rev Respir Dis 1973;108:475-81.
8. Azarow KS, Pearl RH, Hoffman MA, Zurcher R, Edwards FH, Cohen AJ. Vascular ring: does magnetic resonance imaging replace angiography? Ann Thorac Surg 1992;53:882-5.
9. Greenholz SK, Karrer FM, Lilly JR. Contemporary surgery of tracheomalacia. J Pediatr Surg 1986;21:511-4.
10. Applebaum H, Woolley MM. Pericardial flap aortopexy for tracheomalacia. J Pediatr Surg 1990;25:30-2.
11. Hartyanszky IL, Lozsadi K, Marcsek P, Huttl T, Sapi E, Kovacs AB. Congenital vascular rings: surgical management of 111 cases. Eur J Cardiothorac Surg 1989;3:250-4.
12. Roesler M, de Leval M, Chrispin A, Stark J. Surgical management of vascular ring. Ann Surg 1983;197:139-46.