A Radiological Chronicle of the Presentation and Management of a Long Gap Oesophageal Atresia

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ABSTRACT

Long gap oesophageal atresia is a clinically and technically challenging condition to manage. Documentation of the gap between the upper and lower pouches is critical to deciding the timing and feasibility of a primary anastamosis. Integral to this process is the role of radiology. We present a case of long gap oesophageal atresia accompanied by chronological radiography demonstrating its' staged management and highlighting some common complications.

CASE REPORT

INTRODUCTION

Oesophageal atresia (OA) is a rare congenital anomaly with an incidence of 1 in 2500(1). The outcome is dependent on associated anomolies and the length of the gap between the upper and lower pouches of the oesophagus (2,3). Long gap oesophageal atresia (LGOA) is defined as when the upper and lower pouch cannot be easily brought together, and is generally accepted as a distance of between 2 and 6 vertebral bodies (1). It is a rare extreme of OA which has an incidence of approximately 2- 3 per 10,000 live births (4,5). LGOA is technically challenging. Successful delayed repair or oesophageal replacement have significant morbidity (3,6,7). Despite improvement in overall survival, (8) mortality is between 27% and 35% (3,6). Reported is a case of a term infant born with long-gap oesophageal atresia without tracheo-oesophageal fistula and highlighted is the role of radiology in diagnosis and final management.

Oesophageal atresia is classified according to the presence of a tracheo-oesophagela fistula (TOF) and the relation of that fistula to the oesophagus. The most accepted used classification in most paediatric surgical texts are: Type A (9%) pure OA without a TOF, Type B (1%) OA with proximal TOF, Type C (82%) OA with distal TOF, Type D (2%) OA with distal and proximal OA, Type E (6%) H-Type fistula intact oesophagus

CASE REPORT

A term female infant weighing 3,250 grams was born in our institution following an antenatal diagnosis of absence of gastric bubble with polyhydramnios. The infant was born in good condition, with an Apgar score of 7 & 9 at 1 and 5 minutes respectively. A working diagnosis of oesophageal atresia was assumed and following stabilisation on neonatal intensive care, with placement of a replogle tube, a plain chest and abdominal film was performed. This demonstrated a gasless abdomen with a nasogastric tube in the mid-oesophagus (figure 1). The initial film raised some concern that the naso-gastric tube may have perforated the upper pouch

because it was level with the 5th thoracic vertebrae. To address this concern an upper gastro-intestinal contrast study was requested (figure 2) and performed using 2 mls of iopamidol (niopam 300), [Bracco UK, High Wycombe, HP10 0HH], a low osmolar iodinated contrast agent, which clearly showed an intact upper oesophageal pouch to the level of the 5th thoracic vertebrae. The gasless abdomen and oesophageal atresia was suggestive of absence of a tracheo-oesophageal fistula. Further investigative work-up including cardiac, renal and head scans revealed no other congenital anomalies.

Enteral feeding was commenced following a laparoscopic gastrostomy (figure 3). Full enteral feeding was established and remained unremarkable for the first 2 weeks. At 16 days of age, increased biliary discharge from around the gastrostomy site was noticed which prevented enteral feeding. A contrast study through the gastrostomy catheter with iopamidol demonstrated migration of the catheter balloon into the duodenum causing obstruction (Figure 4). The gastrostomy was removed and replaced with a button type device and feeding was re-commenced.

The infant gained weight steadily and contrast studies via the gastrostomy were performed at 26 and 61 days to assess the gap between the upper and lower pouches (figures 5 and 6). These helped us ascertain whether primary closure would be possible or whether a gastric pull-up, gastric tube or intestinal interposition would be required to establish continuity of the oesophagus. The gap, as assessed by radiology, was no more than 3.5 cm's, and with growth of the lower pouch, primary anastamosis of the oesophageal ends was deemed feasible.

A delayed primary oesophageal atresia repair was undertaken at 73 days of age and despite a separation gap of 3.5 to 4.5 cm's between the oesophageal ends, a tension free anastomosis was performed. A contrast swallow 5 days post-operatively clearly demonstrated patency of the oesophagus with the absence of either an anastomotic leak or stricture (figure 7). The infant made an uncomplicated recovery and was discharged home after 79 days of admission. On last review at a few months of age the infant was breast fed and thriving and had no clinical signs of either stricture or significant gastro-oesophageal reflux.

DISCUSSION

Long-gap oesophageal atresia is a technically challenging condition and is reported to occur in 16% of all patients with OA (3). Techniques to reduce the distance between the two ends of the oesophagus for delayed primary repair remain varied and controversial with evidence supporting both upper and lower pouch growth (2,6,7,9). Failing a delayed primary anastomosis being possible makes oesophageal replacement surgery usually necessary (1,10,11).

Initial diagnosis of an infant with OA is suspected antenatally with the absence of a gastric bubble. This is confirmed at birth when a nasogastric tube is unable to be passed into the stomach. An initial babygram usually demonstrates a naso-gastric tube within the cervical oesophagus. The presence of air within the intestine signifies the presence of a tracheo-oesophageal fistula (TOF), although an x-ray should be taken atleast 4 hours post delivery to confirm this (12). These patients require urgent surgery to ligate the fistula. A thoracotomy is performed and at the time of ligation an assessment is made as to whether a primary anastomosis is possible. In the absence of a TOF the initial operation is gastrostomy placement. In infants born with pure OA it is not possible to determine the distance between the upper in lower pouch until a contrast study via a gastrostomy is perfomed.

In this case the initial babygram demonstrated a nasogastric tube at the level of the carina. A known complication of passing a replogle tube is oesophageal perforation (13,14), which in this case necessitated a contrast swallow to exclude this possibility.

Exclusive gastrostomy feeding in infants is commonly associated with leaking around the gastrstomy site. When there is a sudden increase in gastrostomy leakage a contrast study should be performed to exclude catheter balloon migration(15) with consequent gastric outlet or duodenal obstruction. Large bile aspirates through the gastrostomy is indicative of the latter and was seen in this infant and is clearly demonstrated in figure 4. This is more likely to occur with a Foley type gastrostomy catheter with a balloon at its tip which has a predisposition to be propelled by peristaltic activity within the gut. Failure to recognise this may result in impaction and erosion of the wall with bowel perforation. However, prompt decompression of the balloon with withdrawal of the catheter averts this major complication. In addition change from Foley to a button type gastrostomy device which is held against the anterior gastric wall guards against a recurrence.

It is not known to what extent pouch growth is possible in long gap oesophageal atresia. Spitz comments that in long gap oesophageal atresia with a gap of greater than 6 vertebral bodies the chances of saving the oesophagus are remote (1). Various techniques have been proposed to aid this process including internal and external traction of the oesophageal ends (16,17). Over feeding via the gastrostomy to encourage oesophageal reflux is thought to stimulate lower pouch growth. In patients with pure oesophageal atresia, a reduction in the gap between the upper and lower pouches are seen whilst waiting for a delayed primary repair which is usually timed for 8 to 10 weeks post diagnosis(6). From this case report there is objective evidence of this in figures 5 and 6.

Long term outcome in a similar group of patients with pure OA showed 21/27 children underwent delayed successful OA repair. Mortality was 19% in this group and 6 required further resection of the oesophagus, 10 required repeat dilatations and 9 required fundoplication (6).

Following delayed primary repair of LGOA the risk of anastomotic breakdown and stricture is high with many patients requiring repeat oesophageal dilatations or re-resection of oesophagus (2,6). In this case we were able to

perform a tension free repair and no stricture or anastomotic leak was seen on contrast study five days post-operatively.

Furthermore feeding milestones after initial delay are often normal even in LGOA (18) and this is the expectation for this case.

TEACHING POINT

Radiology is critically important in the initial diagnosis and ongoing management of long-gap oesophageal atresia. Contrast studies are useful for measuring the growth of the lower pouch and in assessing the distance between the upper and lower oesophageal pouches. Following a period of waiting (up to 10 weeks) there is growth of the lower pouch which facilitates a primary anastamosis of the ends. Feeding into the stomach via the gastrostomy assists development of the lower pouch.

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FIGURES

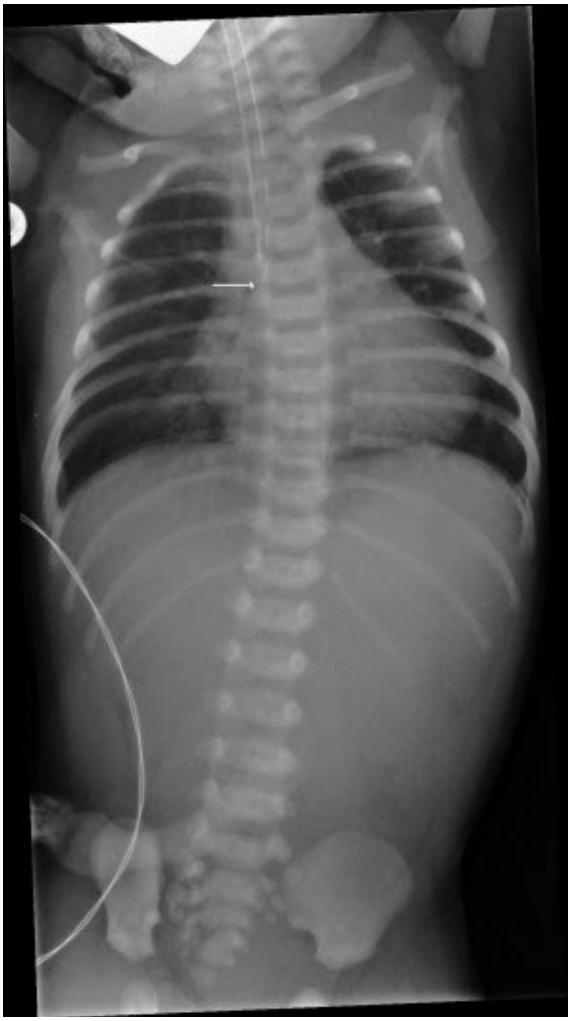

Figure 1: Day 1 babygram of female patient with suspected oesophageal atresia (OA): demonstrating a gasless abdomen with tip of nasogastric tube (arrowed) at mid-thorax level. This is lower than would be expected in a classic OA and a perforation of the upper oesophageal pouch was suspected.

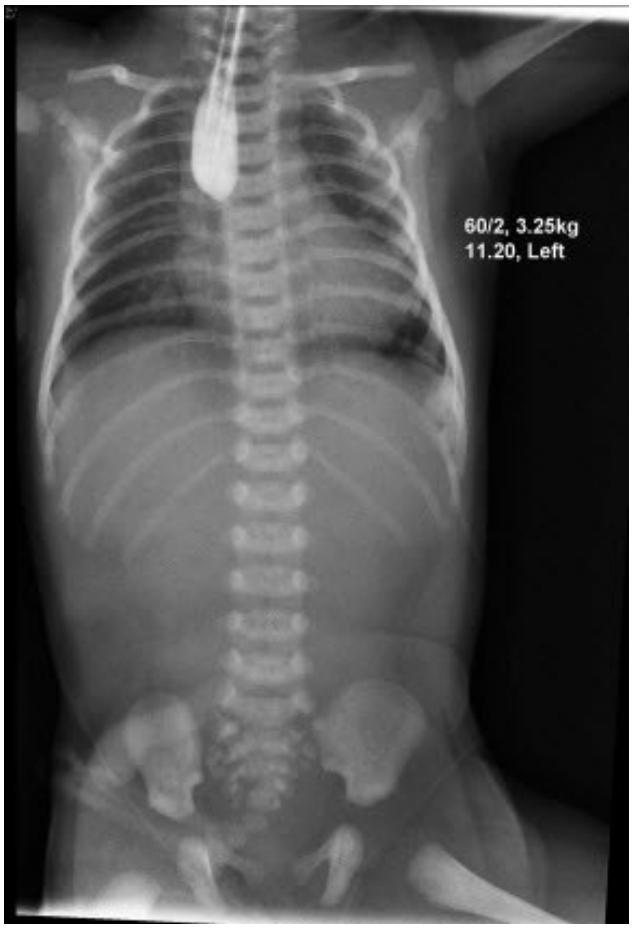

Figure 2: Female patient with suspected oesophageal atresia at 12 hours of age: Contrast study with iopamidol demonstrating a well developed and intact upper oesophageal pouch extending to mid-thoracic level.

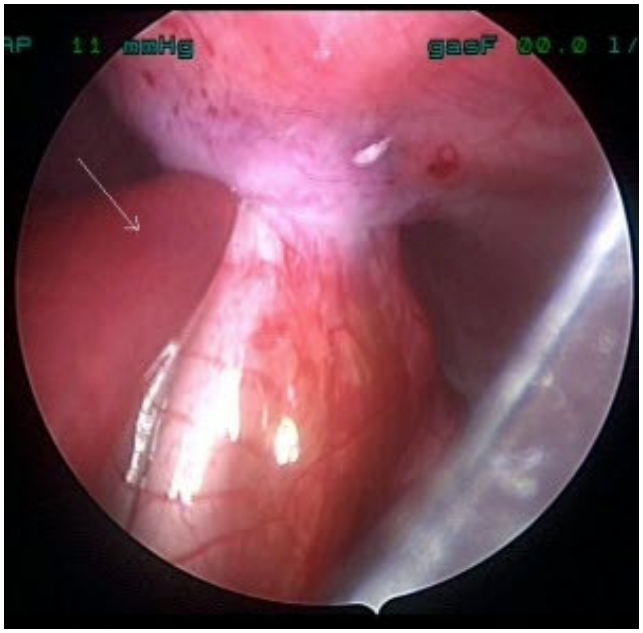

Figure 3: One day old female patient with oesophageal atresia: Laparoscopic gastrostomy creation. Intra operative snapshot demonstrates under developed stomach drawn to anterior abdominal wall by gastrostomy tube, with inflated balloon at tip, in-situ. The liver (arrowed) lies to the left of the stomach.

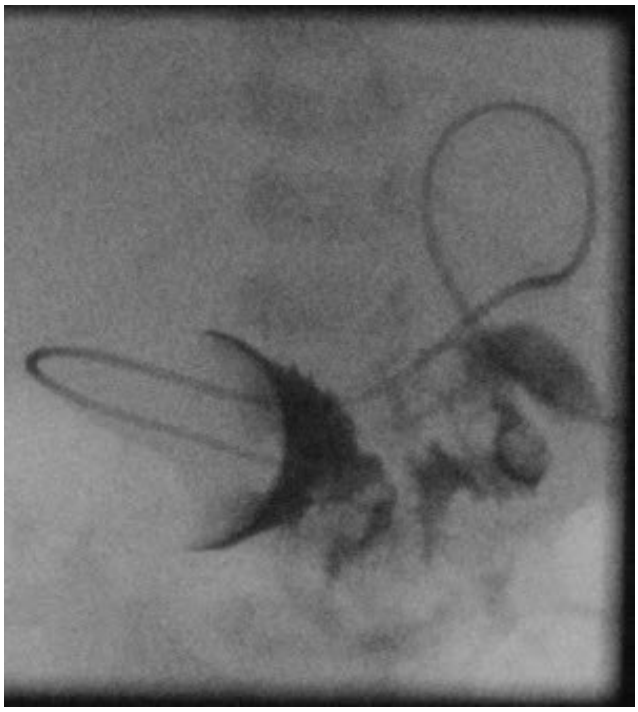

Figure 4: Eighteen day old female patient with oesophageal atresia: Contrast study showing gastrostomy tube migration into distal duodenum with catheter balloon obstruction of lumen. The clear outline of an inflated balloon can be seen as can contrast filling of the proximal jejunum.

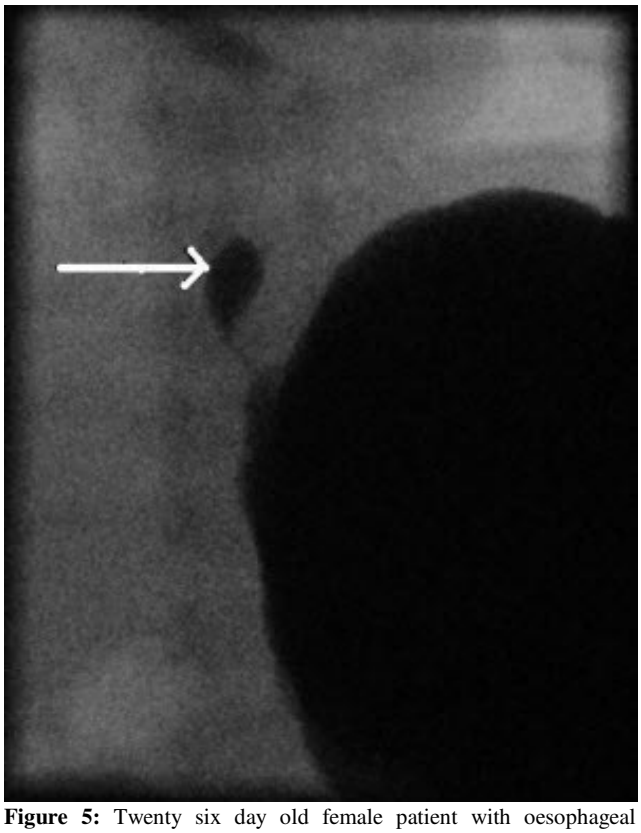

Figure 5: Twenty six day old female patient with oesophageal atresia: Contrast study performed via gastrostomy demonstrating growth of stomach but with a rudimentary lower pouch of oesophagus (arrowed) extending no proximal than level of diaphragm. The upper pouch was not investigated during this study.

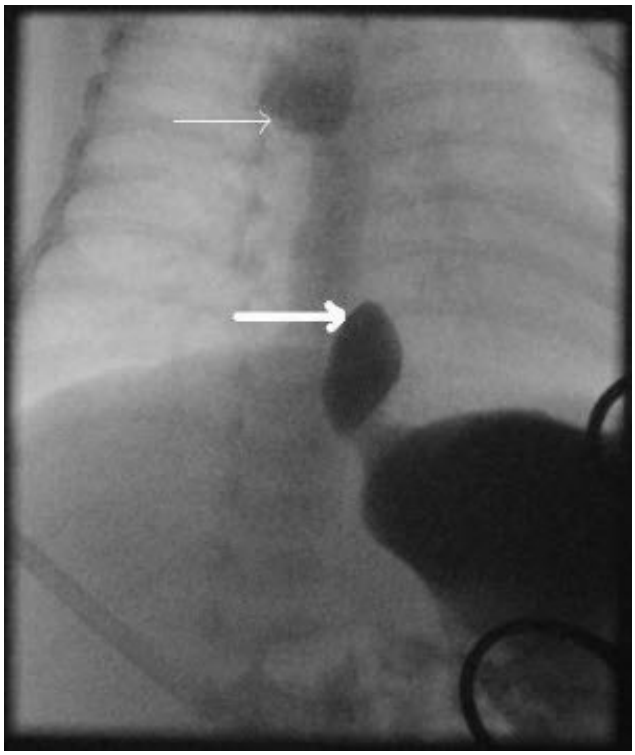

Figure 6: Sixty one day old female patient with oesophageal atresia: Contrast instillation into upper pouch (fine arrow) and stomach demonstrating growth of the lower pouch and a narrowing gap. The separation is estimated to be approximately 3.5 cm's and pouch growth 1 ½ to 2 vertebral bodies. Lower pouch (wide arrow) is noticeably larger than compared to figure 5.

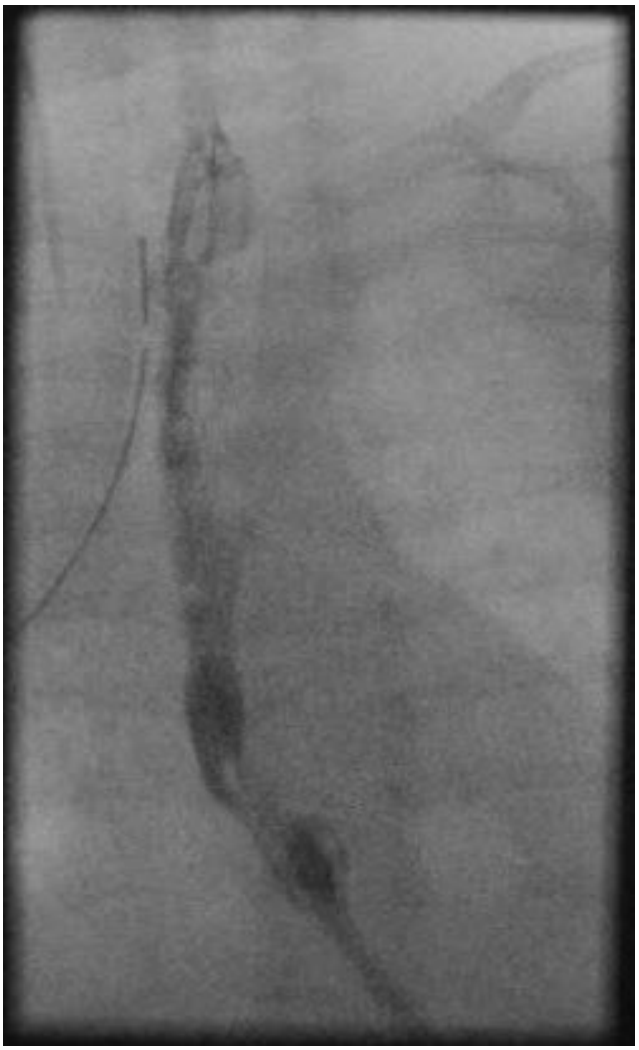

Figure 7: Seventy seven day old female patient with oesophageal atresia: Contrast swallow performed 5 days after primary repair of oesophageal atresia excluding leak or stricture at the anastamotic site which is estimated to be in the distal third of the oesophagus. The intercostal drain which is placed at the time of surgery is seen lying to the right of the reconstituted oesophagus.

ABBREVIATIONS

OA: Oesophageal atresia
LGOA: Long gap oesophageal atresia
TOF: Tracheo-oesophageal fistula.
CXR: Chest X ray

KEYWORDS

Long gap oesophageal atresia, delayed primary repair, infant

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