

CASE REPORT

Surgical Surprise in a Case of Pure esophageal atresia: How High Can the Lower Pouch Be?

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ABSTRACT

The gap between the upper and lower pouches in case of pure esophageal atresia (EA) is usually long and requires multistaged repair. However, in rare cases, the gap may be short and can be repaired primarily commonly through thoracotomy approach. We report an unusual case of a 2-day-old baby girl who presented with short gap EA where the lower esophageal pouch was found adjacent to the blind upper pouch in the neck and primary repair could be performed through the cervical route itself.

Key words: Esophageal atresia; Primary repair; Short gap; Tracheoesophageal fistula

INTRODUCTION

Esophageal atresia (EA) with tracheoesophageal fistula (TEF) is a congenital anomaly occurring in 1 in 2500–4000 newborns [1]. Pure EA accounts for 8–10% of all TEF [2]. In pure EA, the gap between the upper pouch and the lower pouch is usually long. To bridge these long gaps, multistaged surgeries or techniques such as the Foker's, Scharli's, Kimura's, bougienage, traction and delayed anastomosis, and esophageal replacements have been described [3-5]. The native esophagus, however, remains the best conduit. This can be preserved in cases of short-gap EA (SGEA) which can be repaired primarily by the thoracic route. We describe a case of SGEA where the location of the lower esophageal pouch was found as high-up as in the neck, and the repair was done through the cervical route itself. To the best of our knowledge, no such case has been reported earlier.

CASE REPORT

A 2-day-old baby girl with a history of frothing of saliva from the mouth since birth, and inability to feed was referred to us from a peripheral hospital. The child was delivered at 35 weeks by a normal vaginal delivery to a 23-year-old primigravida. When the child presented to us, there was no antenatal ultra-

sound report available, but the mother gave a history of excessive amniotic fluid during the past trimester. The mother also gave a history of attempting to feed the child before coming to our hospital. The baby weighed 1.9 kg and her general condition was sick, dehydrated, heart rate 150 beats/min, peripheries were cold, and capillary refill time was delayed. Saliva was drooling from her mouth. A red rubber catheter inserted perorally failed to go beyond 10 cm. Other systemic examination was normal.

X-ray revealed coiling of an oral catheter in the upper pouch and a gasless abdomen. The upper pouch could be seen ending at the third thoracic vertebral level. No other cardiac, skeletal, or limb abnormalities could be appreciated. The provisional diagnosis was pure EA.

After adequate resuscitation, the neonate was taken up for surgery (cervical esophagostomy and gastrotomy). Assuming it to be the usual long-gap EA, a right cervical skin crease incision was given first for a cervical esophagostomy. After raising subplatysmal flaps, the deep cervical fascia was opened. The carotid sheath and its containing structures were retracted laterally, and the upper esophageal pouch was sought for. A tubular structure, other than the trachea was identified. When traced distally, it was continuing into the thoracic cavity. On meticulous dissection proxi-

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mally, the tubular structure surprisingly ended as a blind pouch. To our surprise, we found another thick-walled dilated tubular structure ending as a blind pouch just above the previous one. The peroral red rubber catheter was found to be located in this, confirming it to be the upper pouch. The gap between the two ends was less than a centimeter. Both the presumed upper and lower pouches were opened (Figure 1). An infant feeding tube (IFT) could easily be passed into the stomach from the open end of the lower pouch. Air was pushed through IFT and its position was confirmed. X-ray done intra-operatively showed the presence of gas in the distal bowel. The upper pouch was adequately mobilized for 1–1.5 cm for a tension-free anastomosis. Any associated upper pouch fistula was ruled out. The two ends of the esophagus were anastomosed primarily over a 6F IFT with 5.0 polyglactin sutures in single-layer full-thickness interrupted fashion from the cervical route itself (Figure 2) in 50 min with minimal blood loss.



Figure 1: The blind-ending upper esophageal pouch and the upper end of lower esophageal pouch seen adjacent to each other through the cervical incision



Figure 2: End-to-end tension-free primary anastomosis of both the ends of esophagus performed through cervical route

The baby was discharged on full oral feeds on day 12. The ultrasound of the urinary system and the echocardiography were normal. On follow-up, she was accepting well oral feeds, gaining weight, and an oral contrast dye study done 6 weeks later showed patent cervical anastomosis with no leak/collection/stricture or narrowing at the anastomotic site.

DISCUSSION

Pure EA is usually characterized by a wide gap between the two pouches [2] and a gasless abdomen on X-ray. The other differential diagnosis of gasless abdomen could be Type B EA-TEF which is associated with an upper pouch fistula and is present in <1% of the cases [2]. In pure EA, the upper pouch is thick and muscular like in this case, unlike a thin and flimsy upper pouch seen in Type B EA-TEF. Loosbroek et al. [6] reported a case of EA that presented in a similar manner, upper pouch with a gasless abdomen. However, surgical exploration through thoracotomy revealed an externally normal looking esophagus with internally obstructing membranes at 2 levels. Thus, in pure EA, the gap between the upper and the lower pouch is usually wide (cannot easily be repaired primarily), but sometimes it may be short [7] or even absent [6]. In most cases of wide-gap EA, a cervical esophagostomy and a gastrostomy are the preferred initial surgery, followed by replacement later on. In this case too, we presumed that the gap between the upper and lower pouches would be wide and hence went in for cervical esophagostomy first.

Primary repair in most cases of SGEA is achieved through thoracotomy [3-5,7,8]. This case is different as the upper end of the lower esophageal pouch was found in the neck itself. The short gap between the two pouches and the cervical location of the lower pouch enabled an end-to-end primary tension-free anastomosis to be done. This saved on the operative time and total duration of general anesthesia and hence ensured subsequent speedy post-operative recovery. A thoracic approach in this case could have been disadvantageous both to the child and the surgeon. This report stresses that the lower pouch in EA may rarely present as high-up as in the neck. Furthermore, not only the gap between the two pouches but also the location of the upper end of the lower pouch determines the surgical approach and the feasibility of primary repair in EA. Although this single-case report does not change the usual management of a case of suspected EA without fistula on an X-ray, it certainly adds to the knowledge of the existence of the cervical location of the lower pouch lest it could be missed.

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