

Pure Oesophageal Atresia at King Abdulaziz University Hospital from 1992 to 2008G: A Case Series

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Abstract. There are different acceptable methods in the management of pure esophageal atresia in common practice. All of them are staged procedures, like esophageal replacement, delayed anastomosis and elongation methods. The objective of this study was to evaluate the outcome of children with pure esophageal atresia, managed by delayed primary anastomosis & post operative complications in relation to this operative technique. A case series retrospective record was based on this study. All medical records were reviewed of patients diagnosed to have pure esophageal atresia from January 1992 to December 2008G at King Abdulaziz University Hospital, Jeddah, Saudi Arabia. Among ten infants found, eight were treated by delayed primary anastomosis. The waiting period ranged from 1 to 5 months in all patients, except one. A leak was seen in 4 cases, two of them had Livaditis' circular myotomy. Incidence of stricture was (62.5%). Pure esophageal atresia in King Abdulaziz University Hospital makes up 15.87% of all esophageal congenital anomalies. Waterston's classification did not affect the post operative procedure-related complications. Stricture was frequently seen in cases that had Livaditis' circular myotomy, but it responded to dilatation better than other cases anastomosed under tension. Radiologic assessment and the measurement of the gap in terms of vertebral bodies is a practical and reliable method to predict the feasibility of primary anastomosis.

Keywords: Pure oesophageal atresia, Isolated oesophageal atresia, Oesophageal anomalies, Tracheoesophageal fistula.

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Accented for publication: 15 March 2010. Received: 30 June 2009.

Introduction

Oesophageal atresia (OA) is one of the most serious and uncommon congenital anomalies that presents at birth; it should be diagnosed early and treated promptly to have minimal mortality and morbidity. Pure OA (isolated atresia) was diagnosed by coiled nasoesophageal tube and gasless abdomen on the chest, plus an abdomen X-ray. It constitutes 6-15% of all congenital esophageal anomalies in different studies. Different surgical treatment plans are available in denoting that no single ideal method was universally accepted.

Method

This is a case series retrospective record based study. All medical records of patients with diagnosis of OA without tracheoesophageal fistula (TEF) seen at King Abdulaziz University Hospital (KAUH), Jeddah, Saudi Arabia, between 1992 and 2008 inclusive were reviewed. Only the patients treated by the management protocol mentioned below were included. Retrospective analysis for demographic data, preoperative, operative and post operative details and follow-up results were presented.

Management Protocol

Once the case was diagnosed as OA without TEF, then a nasoesophageal pouch radio labeled double lumen tube was inserted and kept under continuous low suction. This offered all necessary neonatal care and investigations. Once a patient has been stabilized, mini Laparotomy was done, and double purse string absorbable stitch was applied on the anterior surface of the stomach. A small caliber Hegar anal dilator was introduced through the stomach purse string trans-hiatally into the lower esophageal pouch. At the same time, the anesthetist was asked to gently push-in the already present esophageal pouch tube. A portable X-ray film documented the gap between upper and lower esophageal pouches. If the gap was two vertebrae or less, the definitive anastomosis was done through right thoracotomy with extra pleural approach; after finishing tube gastrostomy operation. However, if the gap is more than two vertebrae, which is commonly the case, then the patient was nursed in ICU or in a special care ward and fed via gastrostomy. The upper pouch

was kept empty by continuous or intermittent suction. Gap measurement was repeated in the same way, but in the X-ray Department every 3-4 weeks.

Results

A total of 10 out of 63 cases were found on congenital esophageal anomalies reviewed. 10 cases were seen of pure atresia, making a ratio of 15.87%. Five cases (50%) were delivered by spontaneous vaginal delivery (SVD), and 5 cases (50%) were preterm 34-37 week (mean 35.4 weeks). Thus, the male to female ratio was of 5/3. Three cases (30%) were delivered at the center. Birth weight ranged from 1.87–3.3 kg (mean 2.26 kg). APGAR score was at 1 min, ranged from 2-8 (mean 6) and was from 7-10 (mean 9) at 5 min. Seven cases (70%) were presented with drooling. Three cases (30%) were presented with cyanosis. Only one patient did not have any other congenital anomalies. All of the others had one or more of the following: chromosomal, cardiac, renal, ano-rectal genital or skeletal anomalies. When applied Waterston classification, there was one case of Class A, 5 cases of Class B and 4 cases of Class C. All patients were diagnosed by chest X-ray which showed coiled naso-esophageal tube in the upper esophageal pouch and gasless abdomen. Four of them had preoperative contrast study before referral to our centre, which supported the diagnosis. Eight cases (80%) underwent extra pleural approach. Five of the eight cases had Livaditis circular myotomy to overcome the gap between esophageal ends (Table 1). Post operative ventilation was needed for 5-7 days (mean 6 days). Start of naso-gastric feeding was performed at 3-9 days (mean 4.57 days). Oral feeding was tolerated between 8-42 days (mean 19.8 days). The incidence of failure to perform primary anastomosis was nil. Stricture was seen in 5 cases (50%), all had dilatations from 1-5 times (mean 3times). Leak was seen in 4 cases (40%), two of them had Livaditis myotomy. Hospitalization period ranged from 12 to 170 days (mean 81 days). Last follow up age was 14 month – 16 years (mean 3.8).

In the remaining 2 cases, no thoracotomy was done due to the severity of associated congenital anomalies. Two patients died in the course of study. The first case was a male, born 2.14 kg, at 38 wks, with Edwards Syndrome; no operation was done for him; died during the 1st 24 hours of life. The second case was a male, 35 wks, BW 1.8 kg with

VACTERL Anomalies (Esophageal atresia, congenital heart disease, ectopic right kidney, imperforate anus and undescended testis); no thoracotomy was done for him, only gastrostomy and colostomy at the age of 2 days. Later, he developed septic shock and died at the age of 38 days.

Table 1. Clinical information of surviving patients.

No.	Sex	Term	BW	Congenital Anomaly	Age at Op	Vent.	Leak	Fed/Oral	Follow-up months	Myotomy	Dil
1	M	35	2.5	CHD, Imp Anus	86 D	6 D	Yes	35 D	35, S	Yes	Nil
2	M	34	2.05	CHD	92 D	7D	Yes	42 D	37, S	Yes	2
3	M	34	2.53	Dysmorphism	32 D	5 D	Yes	20 D	4, S	Yes	Nil
4	M	35	2.8	Down's Syndrome	420 D	6 D	No	9 D	28, D	No	4
5	F	FT	1.87	IUGR	67 D	No	No	9 D	192, D	No	11
6	F	FT	3.02	Dwarfism	100 D	No	Yes	6 D	9, S	Yes	2
7	F	FT	2.17	Down's / PDA	152 D	7 D	No	23 D	5, S	Yes	Nil
8	M	FT	NA	Nil	149 D	No	No	8 D	12, S	No	1

M = Male; F = Female; FT = Full term; CHD = Congenital Heart Disease; IUGR = Intrauterine Growth Retardation; S = Satisfactory; D = Dysphagia; Dil = Dilatation

Discussion

There are different acceptable methods of the management for long gap pure esophageal atresia in common practice. All of them are staged procedures, like esophageal replacement using different gastro intestinal tract, parts as conduit *e.g.* gastric pull up, gastric tube, small intestine, ileo-cecal segment or colon^[1-3]. Plus other methods like elongation procedure or delayed primary repair. It is believe by almost all pediatric surgeons that the best esophageal conduit is the native esophagus itself. This led to the use of intra operative elongation method of the upper esophageal pouch. And occasionally, also the lower one, by circular myotomy, or even suturing them under maximal tension^[4-6]. Delayed primary esophageal anastomosis without preoperative lengthening procedure, described in 1982 by Puri *et al.*^[7], awaiting spontaneous elongation, gained major acceptance worldwide as a procedure that retains and uses the native esophagus in long gap atresia. However, the main problem in this method was the long term hospitalization, as in most cases, it will need about 3 months for the gap to be amenable,

primarily for suturing. The idea of different lengthening procedures was initiated in order to cut the cost of prolonged hospitalization^[8,9].

Anastomotic leak was one of the most common complications seen in all different types of esophageal repair^[12-15]. In all 4 cases, complicated with leak in our series two had circular myotomy. The contrast was contained within the peri-esophageal space; three of them were not clinically evident; the 4th one did show saliva in the chest tube, but not in pleural space. Five patients had Livaditis circular myotomy in lower and/or upper pouches at one, two and even three levels in one patient. This was reflected in the gap length, which were 2-4 vertebrae in our patients at the time of delayed primary anastomosis.

Five of our cases developed stricture, which was managed successfully by esophageal dilatation in all of them. The dilatation was needed one to two times in three patients; all of them had circular myotomy during esophageal anastomosis to overcome tension. The other two cases had 4 and 10 dilatations, respectively; both patients had anastomosis done under tension with no circular myotomy. Although it was difficult to draw solid conclusions from this small number, the results were in favor of circular myotomy to decrease resistant stricture development. This was also supported by different authors^[16,17].

Finally, with the major improvement of thoracoscopic instruments and experience, minimally invasive repair became the fashion of this decade^[10,11]. Delayed primary esophageal anastomosis policy was adopted in the treatment of all pure esophageal atresia, and was applied in 10 cases in our centre. The age at anastomosis for seven cases ranged from 32 to 150 days (mean 96 days). One case was operated at the age of 14 months due to late referral.

It was obvious that the two mortalities were based on Waterston's classification Type C and both died before definitive surgery was performed on them.

Conclusions

1. Pure esophageal atresia constituted up to 15.87% of all congenital esophageal anomalies in our centre.
2. Waterston's classification did not affect the post operative procedure-related complications, like stricture or leakage.

3. Stricture was seen more frequently with circular myotomy, but it responded to dilatation better than other cases anastomosed under tension.

4. Radiologic assessment and the measurement of the gap in terms of vertebral bodies were practical and reliable methods to predict the feasibility, as well as timing of delayed primary anastomosis.

5. No failure in performing delayed primary anastomosis among the studied cases.

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حالات الانسداد الخلقى الكامل للمريء بمستشفى جامعة الملك

عبد العزيز من يناير ١٩٩٢ إلى نهاية ديسمبر ٢٠٠٨م

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المستخلص. توجد طرق متعددة ومقبولة لعلاج الانسداد الخلقى الكامل للمريء. كل هذه الطرق هي مرحلية مثل استبدال المريء، وتأخير المفاغرة، وتطويل المريء. إن هدف هذا البحث هو دراسة وتقييم نتائج الأطفال المصابين بمرض الانسداد الخلقى الكامل للمريء والذين تم علاجهم بطريقة المفاغرة الأولية المتأخرة وكذلك دراسة المضاعفات بعد العملية والمتعلقة بنوعية العلاج. تمت دراسة جميع ملفات المرضى المشخصين بهذا المرض والذين تم علاجهم بمستشفى جامعة الملك عبد العزيز بجدة في المملكة العربية السعودية خلال الفترة من يناير ١٩٩٢ إلى نهاية ديسمبر ٢٠٠٨م. وكان عدد المرضى المصابين بهذا المرض هو عشرة فقط، تم علاج ثمانية منهم بطريقة المفاغرة الأولية المتأخرة والتي تمت بعد فترة انتظار تراوحت من ١ إلى ٥ أشهر عدا مريض واحد. حصل تهريب للصبغة في أربعة مرضى، اثنان منهما أجري لهما قطع ليفاديتيس الدائري لعضلة المريء وكانت نسبة التصبيق بعد الجراحة (٦٢,٥٪). واستنتج من هذه الدراسة أن الانسداد الخلقى الكامل للمريء يمثل ١٥,٨٧٪ من إجمالي تشوهات المريء في مركزنا، وإن تقسيم واترسون لا يؤثر على نوعية المضاعفات بعد هذا النوع من العمليات، كما أن التصبيق بعد العملية لوحظ أكثر ترددا في الحالات التي أجري لها قطع دائري لعضلات

المريء (لأفاديتيس) أكثر من التي تمت المفاغرة فيها تحت شد،
كما أن التقييم الشعاعي لطول المريء مقارنة بعدد الفقرات
الظهرية يعتبر طريقة عملية ومناسبة.