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Oesophageal atresia: Diagnosis and prognosis in Dakar, Senegal

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ABSTRACT

Background: Oesophageal atresia is a neonatal emergency surgery whose prognosis has improved significantly in industrialised countries in recent decades. In sub-Saharan Africa, this malformation is still responsible for a high morbidity and mortality. The objective of this study was to analyse the diagnostic difficulties and its impact on the prognosis of this malformation in our work environment. **Patients and Methods:** We conducted a retrospective study over 4 years on 49 patients diagnosed with esophageal atresia in the 2 Paediatric Surgery Departments in Dakar. **Results:** The average age was 4 days (0-10 days), 50% of them had a severe pneumonopathy. The average time of surgical management was 27 h (6-96 h). In the series, we noted 10 preoperative deaths. The average age at surgery was 5.7 days with a range of 1-18 days. The surgery mortality rate is 28 patients (72%) including 4 late deaths. **Conclusion:** The causes of death were mainly sepsis, cardiac decompensation and anastomotic leaks.

Key words: Esophageal atresia, Diagnosis, Prognosis, Sub-Saharan Africa

INTRODUCTION

Oesophageal atresia is a rare malformation with a prevalence of 2.43 per 10,000 births.[1] After surgery, the survival rate is about 90% if there are associated abnormalities and about 100% in isolated forms.[2,3] In sub-Saharan Africa, mortality is still high, raising several questions about the management of oesophageal atresia.[4] The goal of our study was to analyse the diagnostic difficulties and its impact on the prognosis of this malformation.

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PATIENTS AND METHODS

This was a 4-year retrospective study (2010-2013) based on the records of patients born with esophageal atresia. The patient's profile, medical history, physical findings, laboratory and surgical treatment results were analysed.

RESULTS

There were 49 newborns, including 31 boys and 18 girls, with an average age of 4 days (range: 0-10 days) [Figure 1]. The average birth weight (BW) was 2100 g (range: 1000 g - 3200 g). None of the patientss was diagnosed prenatally. Prenatal ultrasound performed in 41 patients showed hydramnios (7) and intrauterine growth delay (4). There were 14 premature deliveries, 4 before 34 weeks and 2 before 30 weeks. One of the patients products of twin pregnancies, the other twin was normal. The main features at presentation were hypersalivation and respiratory distress [Table 1]. Attempted passage of nasogastric tube got arrested at about 8-10 cm in 44 patients. Plain X-rays of the chest and abdomen in all patients showed the level of the proximal oesophageal pouch located at the second, third or fourth thoracic vertebra. Five patients needed trans-tube instillation of gastrografin to confirm the diagnosis and to determine the level of the proximal oesophageal segment. Associated anomalies were dominated by heart defects and anorectal malformation [Table 2]. Twenty four patients had severe pneumonia on

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admission. Surgery closure of the tracheo-oesophageal fistula and primary oesophageal anastomosis) was performed on 39 patients. The average age at surgery was 5.7 days with a range of 1-18 days. Thirty eight patients had type III atresia while one had type I of atresia. Three patients with anorectal anomally had with colostomy at the same sitting. Intra-operative complications included pleura tear (14), rib fractures (2), and one case of death. All patients were managed in the Intensive Care Unit on intravenous crystalloids. Enteral (trans nasogastric tube) feeding is introduced on the 5th day post-operative on average (range: 3 - 8 days) with gradual increase in the rate on the following days. Gastrographin swallow was performed between the 7th and 10th post-operative days with a nasogastric tube in place to rule out the absence of anastomotic leakage and major oesophageal stenosis before starting oral feeding. There were 11 anastomotic leaks including 4 early (first post-operative day), 2 reccurrence of the tracheo-esophageal fistula and 3 oesophageal stenoses. All the patients developed thrombocytopenia with moderate to severe anaemia. The surgery-related mortality rate was 72% (28 patients). The causes of death were mainly sepsis, cardiac decompensation and anastomotic leaks [Figure 2]. Three patients developed severe enterocolitis after introduction of enteral feeding of breast milk with 100% mortality rate. The average follow-up was 22 months (14-40 months).

Table 1: Main symptoms

Symptoms	*n* (%)
Hyper-salivation	34 (69)
Respiratory distress	20 (40)
Bronchial congestion	21 (42)
Regurgitation	12 (24)
Cyanosis	6 (12)
Abdominal bloating	2 (4)

Table 2: Associated anomalies with oesophageal atresia

Associated malformations	*n* (%)
Cardiac	14 (28)
Anorectal malformation	3 (6)
Choanal atresia	2 (4)
Bilateral cryptorchidism	1 (2)
Right inguinal hernia	1 (2)
Craniofacial dysmorphia	1 (2)

DISCUSSION

The diagnosis of oesophageal atresia is often done during prenatal screening nowadays.[4] The progress made in medical imaging field, the monitoring of high-risk pregnancies and biological analyses of amniotic fluids have permitted early diagnosis. The morphological ultrasound in third trimester and sometimes the foetal magnetic resonance imaging (MRI) done by qualified specialists have clearly increased the chances to visualise the absence of the continuity of the oesophagus associated with hydramnios and/or a small or missing stomach. The key is an ultra-early diagnosis (18 weeks of amenorrhea) and hydramnios (a late sign most often occurring after 24 weeks of gestation).[5-7] Diagnosis by prenatal ultrasound relies on indirect signs (hydramnios and/or small or invisible stomach) and direct signs (dilated proximal oesophagus or 'pouch sign').[6,8] The positive predictive value of the combination of these two signs (hydramnios and invisible or small stomach) is low, between 40% and 56%, with many false-positives;[9-11], while the combination (hydramnios and invisible or small stomach) and a dilated proximal esophageal segment ('pouch sign') is between 60% and 100% with 80% and 100% sensitivity.[8,10,12] The MRI improves the sensitivity and specificity of the prenatal screening.[13,14]

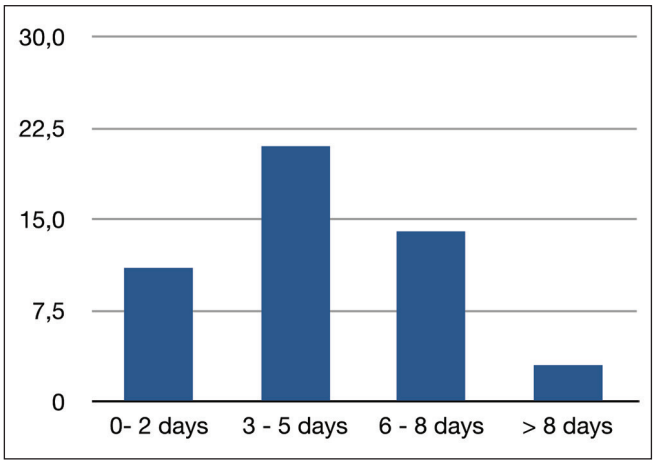

Figure 1: Distribution of patients according to the age of admission

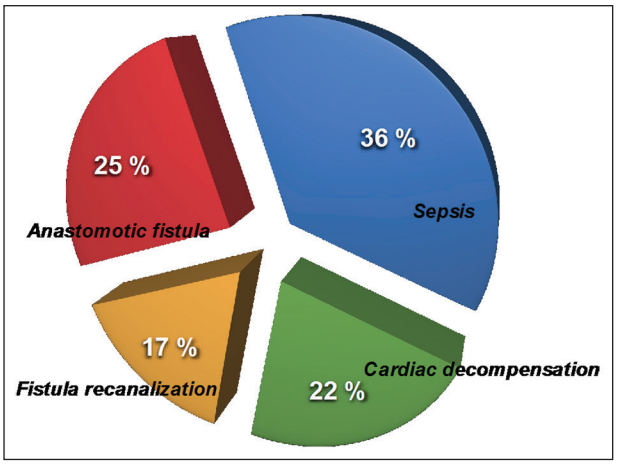

Figure 2: Causes of operative mortality in the short-term

The lack of esophageal continuity induces digestive enzymes changes such as gamma-glutamyl transpeptidase and proteins in amniotic fluid during the 18th week of gestation.[15] These biological changes allow for the diagnosis of oesophageal atresia with 98% sensitivity and 100% specificity, regardless of the anatomic type.[5]

The absence of prenatal diagnosis in our study was due, to a lack of local expertise needed for the monitoring of suspected pregnancies. The hydramnios may have foetal digestive origin in <10%, whereas other causes may be maternal or placental.[16,17] Oesophageal atresias are responsible for a hydramnios in 95% of cases in the absence of fistula and 35% in the presence of distal fistula.[16] The finding of hydramnios should should arouse suspicion necessitating ultrasound from the 22nd week after gestation. Biological analysis of the amniotic fluid is a suitable option to consider. The creation of advanced prenatal screening units and the establishment of multidisciplinary meetings could optimise the management of oesophageal atresia. The prenatal diagnosis allows for in utero transfer of the foetus and the management to a destined birthplace for an expert management.[18]

All of our cases were diagnosed after birth with an average time quite long compared to the literature.[5,18] Often the clinical signs are enough to make the diagnosis; the additional tests are usually needed to establish the type of anatomical variant of oesophageal atresia and associated malformations. Proximity of maternity centers with the Pediatric Medical Surgery facilities, and improvement of collaboration between the various stakeholders who care for mothers and children could significantly shorten the time of diagnosis. The morbidity related to the delay of diagnosis is accentuated by the attempts to feed, thus causing a high rate of preoperative sepsis and pneumonia in our studies. The rate of associated malformation in this study agrees with earlier studies, with a predominance of cardiac abnormalities.[19-22]

Generally, there was a delay in the surgical intervention in this study, largely due to unavailability of preoperative investigations in emergency setting. This is unlike other studies that reported intervention period is of about 48 h.[5,18] The closure of the tracheo-esophageal fistula and primary oesophageal anastomosis is the reference technique with good outcome, as was our experience in this study.[23,24] However, staged operation is advisable in long gap defects (>3 vertebrae), while waiting for the spontaneous growth of the two oesophageal segments in 6-8 weeks. Corrective surgery can then be done by oesophageal anastomosis or by use of a gastric and intestinal transplant subsequently.[24,25]

Table 3: Mortality according to the classification of Spitz

Classification	Number	Operative death	Non-operative death	Survival
Spitz A (BW >1500 g, without major CD)	29	15	05	09
Spitz B (BW <1500 g or major CD)	15	10	03	02
Spitz C (BW <1500 g and major CD)	5	3	2	0

BW: Birth weight; CD: Cardiac disease

Morbidity in our study was mainly caused by sepsis and anastomotic leaks. Koivusalo *et al.*[26] Reported that the longer the defect, the greater the risks during the anastomosis. This also causes gastro-esophageal reflux and oral feeding difficulties.[26] Other studies reported that the two main factors for survival are BW over 1500 g and the absence of associated cardiac abnormalities [Table 3].[27,28]

In our study, mortality caused by sepsis was high especially in patients with a weight <1500 g and an isolated form of oesophageal atresia. This mortality may be directly related to the delay in diagnosis that increased the risk of severe pneumopathies caused by gastric fluid reflux into the lungs when saliva is inspirated and attempts to feed.

CONCLUSION

Oesophageal atresia is a neonatal emergency and life-threatening. Diagnostic delay increases the preoperative morbidity and makes the post-operation more complicated to manage. Thus, early diagnosis, the time for a surgery and the quality of post-operative intensive care determine the results.

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Conflicts of interest
There are no conflicts of interest.

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