MARRAKECH
جامعة القاضي عياض
UNIVERSITÉ CADI AYYAD

كليـــــــة الطب
والصيدلة - مراكش
FACULTÉ DE MÉDECINE
ET DE PHARMACIE - MARRAKECH

Year 2021 Thesis N° 174

Management of long gap esophageal atresia

THESIS

PRESENTED AND PUBLICLY DEFENDED ON 13/10/2021

BY

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Born on: August 17, 1995 in Marrakech

TO OBTAIN THE DEGREE OF DOCTOR OF MEDICINE

KEYWORDS

Long gap – Esophageal atresia – Management

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ذِي عِلْمٍ عَلِيمٌ

صَدَقَ اللهُ العَظِيمُ

Oath of Hippocrates

As a member of the medical profession:

I solemnly pledge to dedicate my life to the service of humanity;

The health and well-being of my patient will be my first consideration;

I will respect the autonomy and dignity of my patient;

I will maintain the utmost respect for human life;

I will not permit considerations of age, disease or disability, creed, ethnic origin, gender, nationality, political affiliation, race, sexual orientation, social standing or any other factor to intervene between my duty and my patient;

I will respect the secrets that are confided in me, even after the patient has died;

I will practise my profession with conscience and dignity and in accordance with good medical practice;

I will foster the honour and noble traditions of the medical profession;

I will give to my teachers, colleagues, and students the respect and gratitude that is their due;

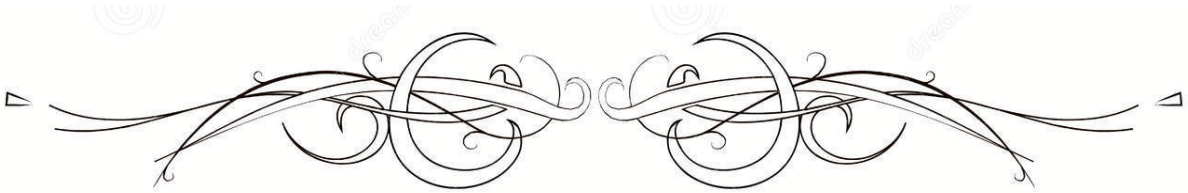
I will share my medical knowledge for the benefit of the patient and the advancement of healthcare;

I will attend to my own health, well-being, and abilities in order to provide care of the highest standard;

I will not use my medical knowledge to violate human rights and civil liberties, even under threat;

I make these promises solemnly, freely, and upon my honour.

Geneva Declaration, 1948



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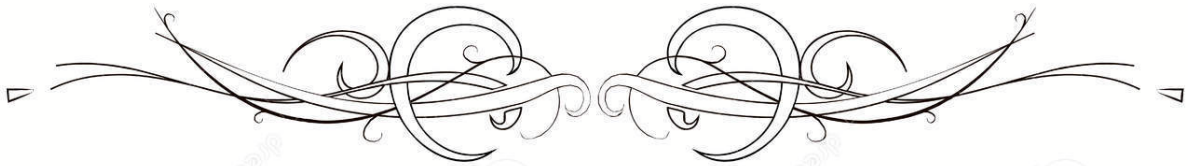

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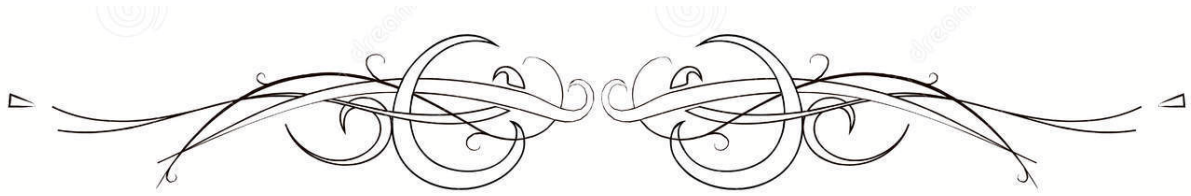
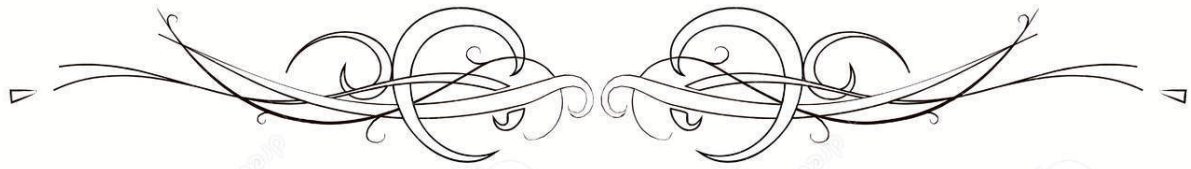


DEDICATIONS



I dedicate this thesis to...

To my beloved mother Malika El Hachimi

Your unconditional love and care mean everything to me. My heart can't thank you enough for giving me such a strong foundation and helping me be the independent adult I am today. Your immeasurable support means so much to me that I really don't know how to repay you. Thank you for all the sacrifices you have made for me and doing everything possible to help me grow and excel in life and for constantly showering me with affection and love. I will forever be grateful for everything, I love you Mom.

To my beloved father Mohamed El Amghari

I'm so much grateful to Almighty Allah, who gifted me a father like you. Your importance in my life could not be expressed in words. Thank you for being a patient listener, for giving me correct advice, for sorting out my problems easily. I'm grateful to you for guiding me to reach new heights and all your words that gave me a perspective which no books can teach me. Whatever I do, wherever life takes me, I will always be thankful to you, I love you Dad.

To my dear sisters Hajar and Salma El Amghari

Having sisters like you is a divine blessing. I would like to thank you for all the times when you were so loving and caring. I do not have words to appreciate you for everything that you do for me. No matter how much we fight and argue at little things, you will remain my darling sisters. Love you.

To my dear paternal family El Amghari and to my dear maternal family El Hachimi

I am so appreciative to have the loyal, supportive, and caring family that I do. Thank you for accepting me for who I am and where I am at right now. It is a huge source of comfort to know how much I am genuinely loved.

In memory of my grandparents

Dear grandparents, you are no longer with us. But your love and memories will always be in my heart forever. May Allah give your souls eternal peace and grant you heaven.

To my brother from another mother Hamza and his family Bensmail

14 years ago I had the honor to meet you. Since then, you have shown me how lucky I am to have you as a brother. I'm so glad I don't know what loneliness means, because you never let me feel lonely. You are my biggest support, my trusted advisor, my secret keeper, my source of joy whenever I am going through hard times. Your family made me feel since day 1 as one of theirs. For that and for many reasons, I want to say thank you. May our friendship be like the infinity loop as it has no ending to it. I can only wish you and your family a happy, healthy, and prosperous life with great success.

To my homy and med school companion Riad El Baroudi

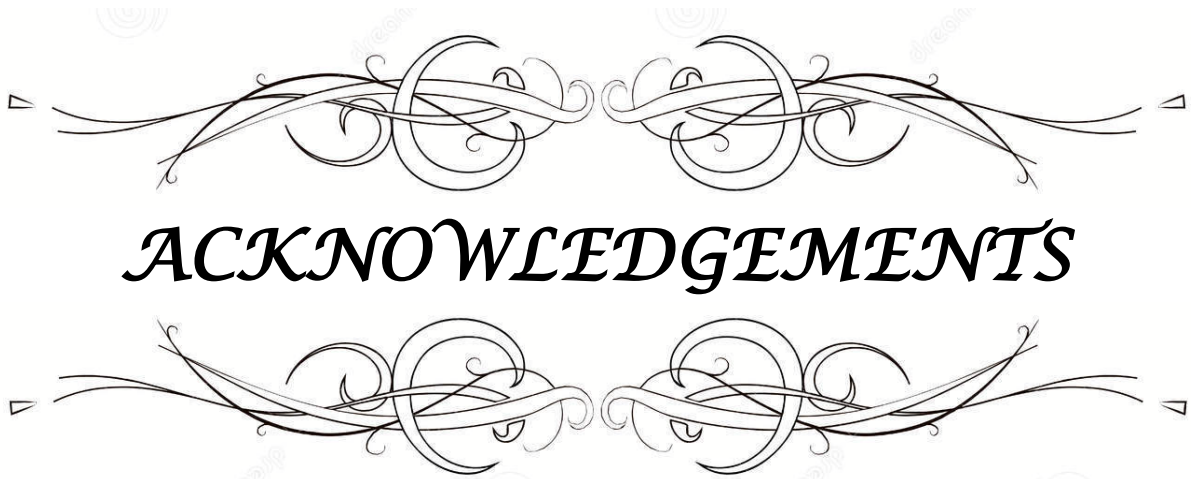
If I could choose the best moment of my time in medical school it would certainly be the day I met you. Your kindness, selflessness, sense of humor have taught me the true definition of friendship. Thank you for always putting a smile on my face when I need it most, for being there to lean on when I need someone, and for being one of the most incredible friends anyone could ever ask for. Please know that I count myself really fortunateto have you in my life as a friend. May our friendship last as long as we're alive and may I be by your side when you become the great doctor you aspire to be, lieber Bruder.

To my rajawi and madridista brother Zakaria Sadak

Behind that teasing and pesky person, lies a great brother with a big heart. It means the world to me that you are there for me when I need someone to lean onand that's something I can only say about few people in my life. Your friendship has helped me through many times in life, and I feel blessed to have you in my life even though sometimes you're a real pain in the neck. Wishing you a bright and colorful future in which all your dreams come true Bruder.

To my dear friends : Sara Mouhmouh, Sara El Ansari, Mohamed Oubihi, Youssef Houmair, Marouane Slimani, Youness Aarab, Mouad Ezzaighi, Redouane Mouflih, Saad Choukry, Mehdi Kouadssi, Zakaria Dararí

You were someone I don't know before; you were someone I don't expect I will get along with. But it turned out you have given me one of the best friendships ever. Thank you.



ACKNOWLEDGEMENTS

To Professor Said Younous

Head of Pediatric Intesive Care Unit

Professor of Anesthesiology-Resuscitation

You have done me a great honor by kindly accepting to chair my thesis jury. Your great knowledge, your dynamism and your kindness have always aroused in me great esteem. Please find here, dear master, the testimony of my great gratitude and high consideration.

To Professor Mohamed Oulad Saiad

Head of Pediatric General Surgery Department

Professor of Pediatric Surgery

You have given me a great honor by accepting to supervise our thesis. Your recommendations, your competence and your rigorous workcompetence are for me an example to follow.

You have always given me the best welcome despite your professional obligations. I thank you infinitely, dear Master, for having devoted to this work a part of your precious time and for having guided me with rigor andkindness. I am very proud to have learned from you and I hope to have lived up to your expectations.

Please accept, dear master, through this work the assurance of my esteem and of my deep respect.

To Professor Ghizlane Draiss

Professor of Pediatrics

I am very touched by the kindness and spontaneity with which you have accepted to judge our work.

I have a great consideration for your extreme kindness as well as for your professional qualities.

Through this work, please accept, dear Master, the expression of my sincere gratitude and deep respect.

To Professor Monir Bourrous

Head of Pediatric Emergency Department

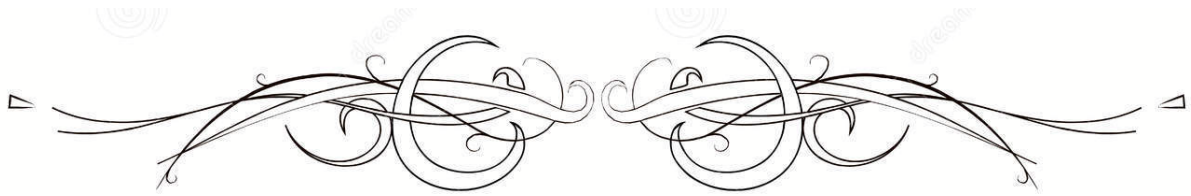
Professor of Pediatrics

You have done me a great honor by accepting to be associated with our thesis jury.

Your undeniable competence, your charisma and your human qualities make you a great professor and inspire me a great admiration.

Allow me, dear master, to express to you

my deep respect and my high consideration.



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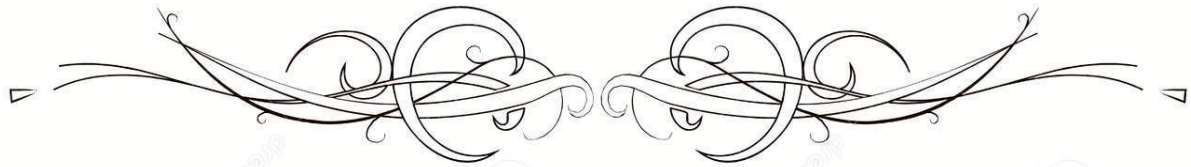


List of abbreviations

ASD	: Atrial septal defect
CBC	: Complete blood count
CHD	: Congenital hip dislocation
CoA	: Coarctation of the aorta
CPAP	: Continous positive airway pressure
CRP	: C-reactive protein
CTE	: Congenital talipes equinovarus
DPE	: Delayed primary anastomosis
EA	: Esophageal atresia
GER	: Gastroesophageal reflux
ICU	: Intensive care unit
LGEA	: Long gap esophageal atresia
MRI	: Magnetic resonance imaging
PDA	: Patent ductus arteriosus
PPI	: Proton pomp inhibitors
PROM	: Preterm rupture of the membranes
RAA	: Right aortic arch
RLD	: Radial longitudinal deficiency
SPSS	: Statistical Package for the Social Sciences
TEF	: Tracheoesophageal fistula
TPL	: Threatened preterm labor
VSD	: Ventral septal defect

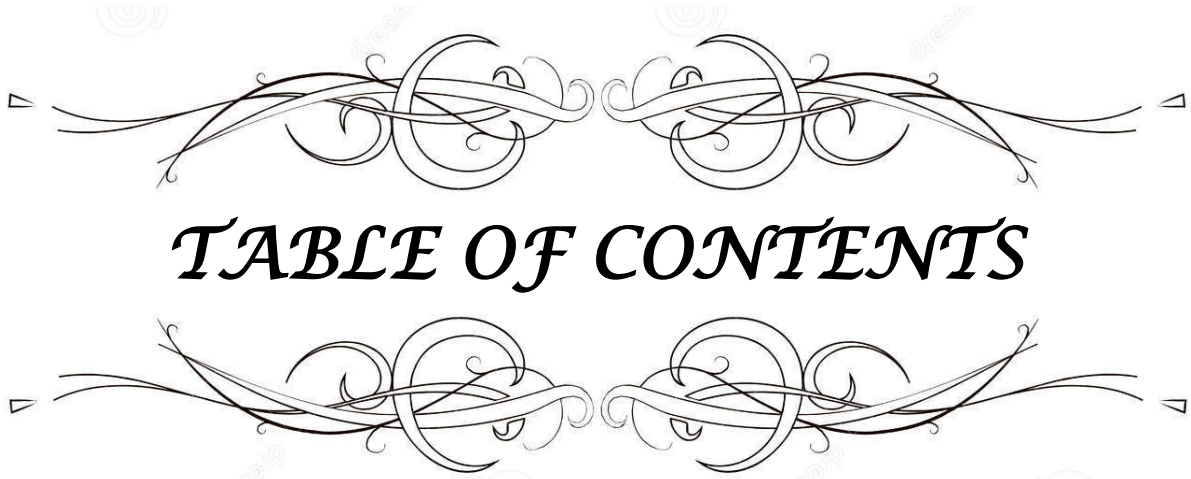
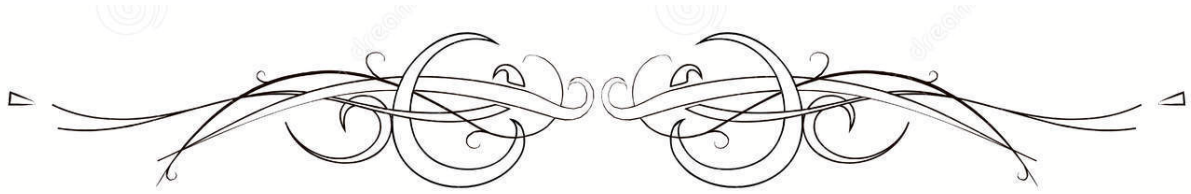
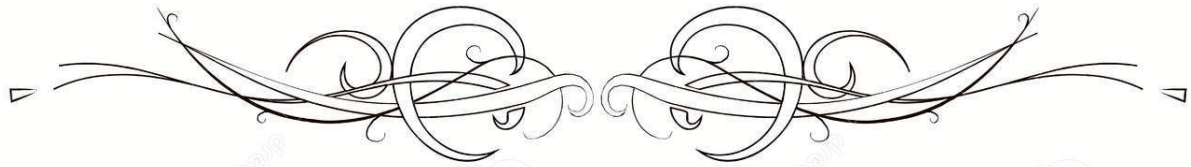


TABLE OF CONTENTS



INTRODUCTION



Esophageal atresia is a birth defect that affects the alimentary tract. It causes the esophagus to end in a blind-ended pouch rather than connecting normally to the stomach. EA is most frequently associated with a tracheoesophageal fistula that represents an abnormal connection in one or more places between the esophagus and the trachea.The estimated prevalence of EA varies worldwide from 1 in 2500 to 1 in 4000 births.[1]

Several classifications have been established for EA, one of which distinguishes long gap EA (LGEA) from non long gap EA, critical in the surgical repair approach. No conventional definition of LGEA has been determined. As there is no standard measurement technique nor specific numerical definition of the gap length between the uper and lower pouch. Some practitioners prefer using vertebral bodies or centimeters to measure the distance between the esophagus ends, while others identify EA as long gap when the primary anastomosis is considered not possible or attempted but failed.

LGEA is associated with high mortality and morbidity, therefore it represents a neonatal surgical emergency and it should be screened during the initial examination of every newborn in the delivery room.

Several surgical techniques with different outcomes have been described for the management of LGEA. However it remains a challenge for the practitioners as there is a lack of consensus regarding the management of this pathology.

The purposes of our study are as follows :

- Report the experience of the General Pediatric Surgery Department of the University Hospital Mohammed VI of Marrakesh, through a series of 21 patients hospitalized in the department between 2013 and 2020.
- Compare the results to the data of medical literature.
- Improve the management of this pathology

MATERIALS AND METHODS

I. Materials

1. Type of study :

This is a retrospective study of a series of 21 patients that was carried out over a period of 7 years, from January 2013 to December 2020.

2. Study framework :

This study was conducted in the General Pediatric Surgery Department and the Pediatric Intensive Care Unit of the University Hospital Center Mohammed VI of Marrakesh.

3. Study sample :

The study included 21 patients diagnosed with esophageal atresia ; 13 patients with pure EA, and 8 EA patients with distal TEF for whom a primary anastomosis was considered impossible intraoperatively.

Patients, who were not operated or for whom a primary anastomosis was performed, were excluded from this study.

II. Methods :

1. Data collection :

For each patient included in the study, the data were collected retrospectively from the medical records and recorded in an operating sheet established for this purpose (annex).

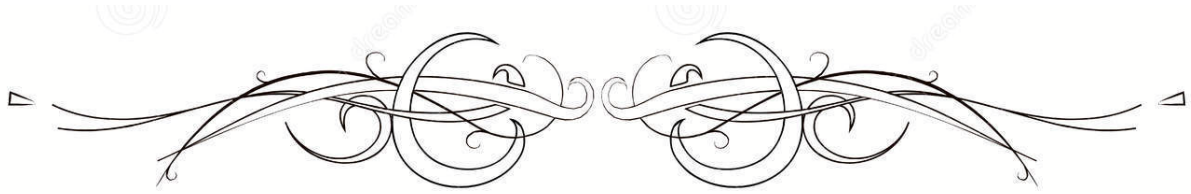
For each medical file included, the following informations have been identified :

- Epidemiological : Pregnancy follow-up, gravidity, parity, consanguinity, history of maternal diseases, familial history of EA, term of the pregnancy, delivery mode, gender, birth weight.
- Clinical : diagnostic delay, functional signs, results of the physical examination.
- Radiographic: position of the nasogastric tube, pulmonary parenchyma, digestive aeration, anatomical type.
- Malformations check-up
- Therapeutic management : management in intensive care unit, type of surgical procedure
- Evolution : Short and long term postoperative complications
- Mortality

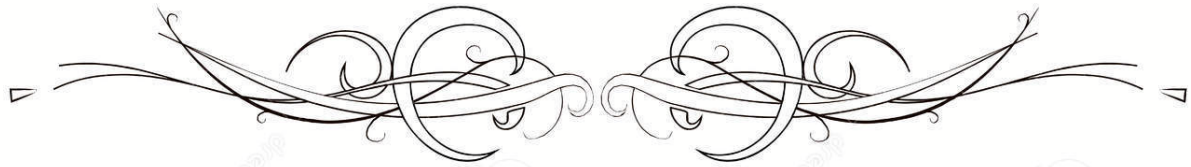
2. Stastical analysis :

The statistical analysis was performed using SPSS Statistics (Statistical Package for the Social Sciences) version 28.0.

Quantitative variables were expressed as mean and extreme values. Qualitative variables were expressed as absolute values and as a percentage.



RESULTS



I. Epidemiological data :

1. Frequency :

134 patients with esophageal atresia were hospitalized in the Department of General Pediatric Surgery during the course of our study. 21 of these 134 patients had a long gap esophageal atresia, representing a percentage of 15.67% of all cases of esophageal atresia and a frequency of 3 cases per year. (Figure 1)

Figure 1 : Annual prevalence of EA and LGEA

2. Pregnancy follow-up :

In our series 18 pregnancies were followed up (86%), while 3 pregnancies were not (14%). (Figure 2)

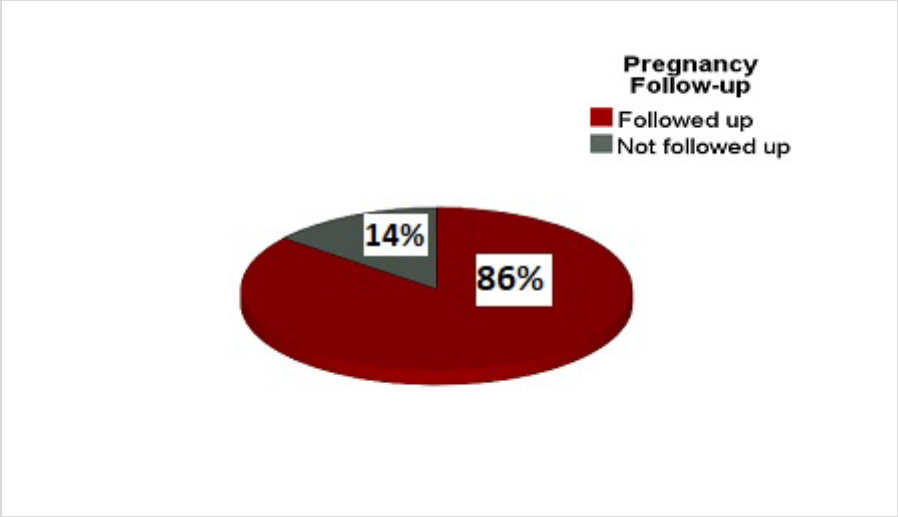


Figure 2 : Distribution of pregnancies by follow-up

3. Course of the pregnancy :

The course of the pregnancy was normal in 12 cases and abnormal in 9 cases. Polyhydramnios was found in 8 cases, premature rupture of membranes (PROM) in 3 cases and threatened preterm labor (TPL) in 1 case. (Table I)

Table I : Observed anomalies during pregnancy

Pregnancy	Number of cases	Percentage
Normal	12	57%
Polyhydramnios	8	38%
PROM	3	14%
TPL	1	4%

4. Parental consanguinity :

Consanguinity was identified in 3 cases (14%), while 18 patients were from a non consanguineous marriage (86%). (Figure 2)

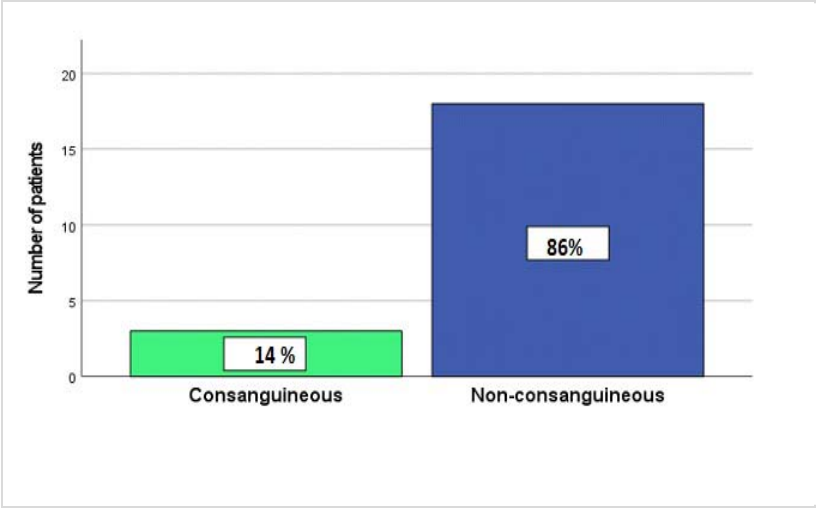


Figure 2 : Rate of the consanguinity

5. Parity :

In our series 11 mothers were primiparous. 10 were multiparous; 4 with 2 parities, 3 with 3 parities, 1 with 4 parities and 2 with 5 parities. (Figure 3)

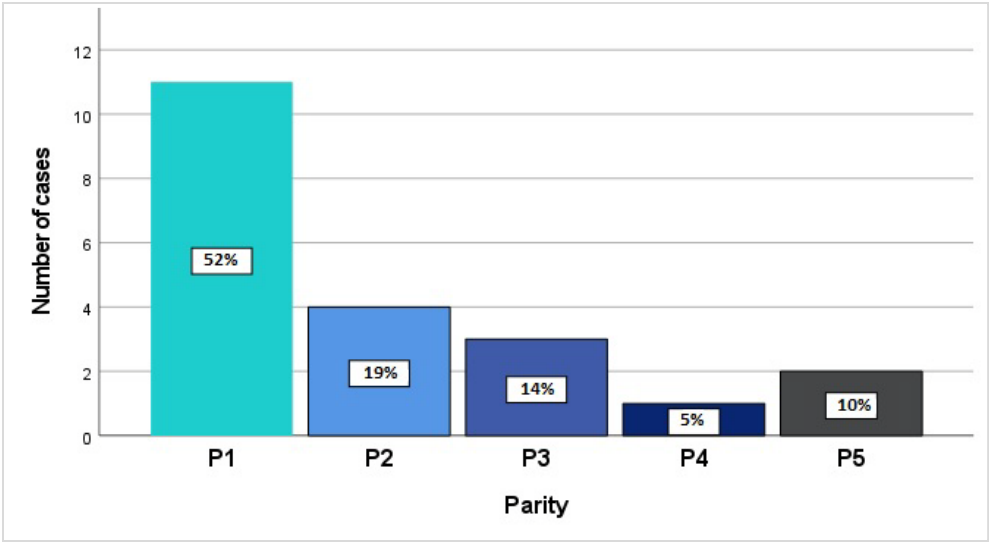


Figure 3 : Distribution of cases by parity

6. Familial medical history of EA :

None of our patients had a familial medical history of EA.

7. Maternal medical history :

No remarkable family medical history was identified in 18 cases. Gestational diabetes was identified and treated with insulin in 2 cases. Systemic lupus erythematosus (SLE) was noted in 1 case. (Table II)

Table II : Prevalence of maternal diseases

Maternal medical history	Number of cases	Percentage
None	18	86%
Gestational diabetes	2	9%
SLE	1	5%

8. Gestational age :

17 newborns were born at term (81%), and 4 were premature (19%). (Figure4)

For preterm newborns, the mean term is 35 weeks (33 weeks + 5 days-36 weeks).

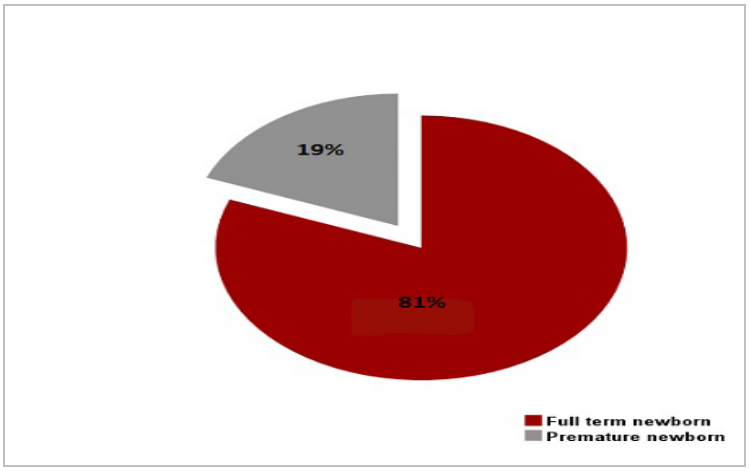


Figure 4 : Distribution of patients by gestational age

9. Delivery mode :

11 patients were born by vaginal delivery, representing a percentage of 52%. And 10 patients were born by cesarean section (48%). (Figure 5)

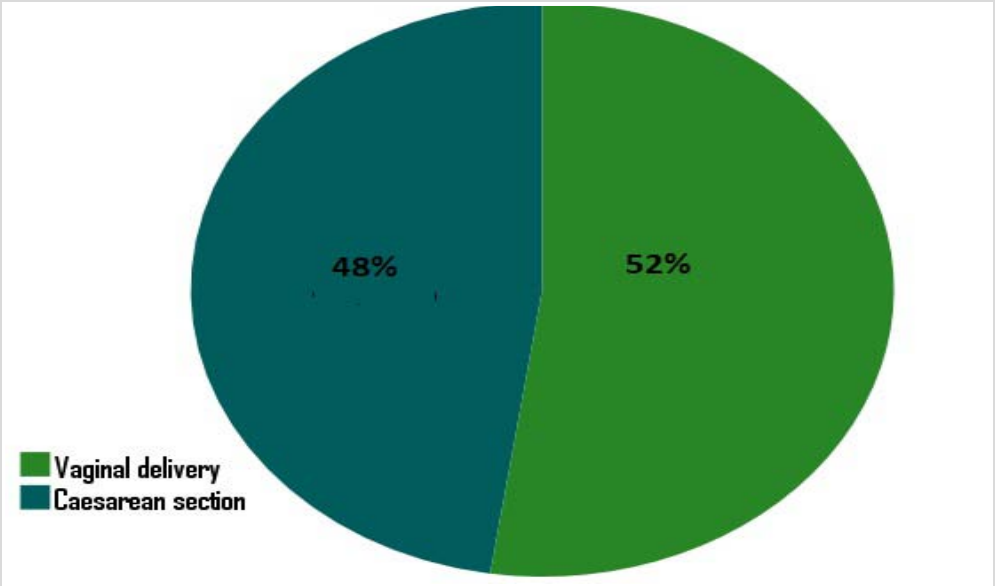


Figure 5 : Distribution of cases by delivery mode

10. Caesarean section indications :

In the 10 performed caesarean sections, 4 parturients had fetal malpresentation as an indication (40%), 3 had an acute fetal distress (30%), 2 had chorioamniotitis (20%), 1 had a previous caesarean section (10%). (Table III)

Table III : Caesarean section indications

Indications	Number of cases	Percentage
Fetal malpresentation	4	40%
Acute fetal distress	3	30%
Chorioamniotitis	2	20%
Previous c-section	1	10%

11. Gender :

Our series included 14 boys (67%) and 7 girls (33%). The gender ratio M/F was 2. (Figure 6)

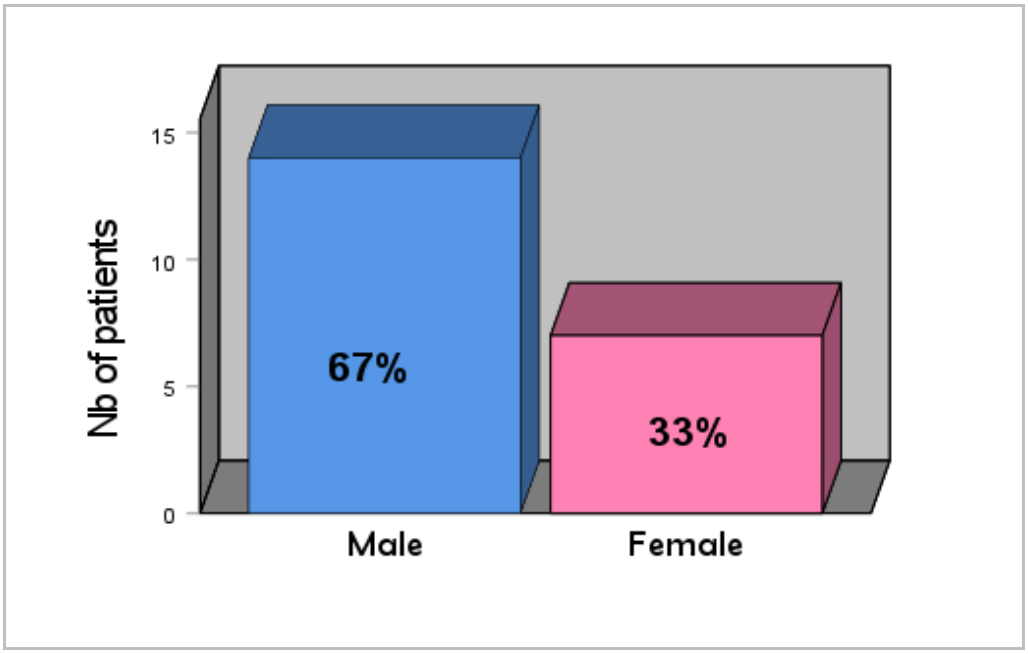


Figure 6 : Distribution of patients by gender

12. Birth weight :

The mean weight of our patients is 2790 grams (2100g-3500g).

14 patients had a birth weight over 2500g (67%), 7 patients had a birth weight between 1800g and 2500g (33%), and none of our patients had a birth weight below 1800g. (Table IV)

Table IV :Patients distribution by birth weight

Weight (W) in grams	Number of patients	Percentage
W>2500g	14	67%
1800g<W<2500g	7	33%
W<1800g	0	0%

II. Clinical data :

1. Diagnostic delay :

In our series we notice :

- 13 patients were diagnosed during the first 24 hours (62%). Among them, 5 were diagnosed immediately after birth following a systematic screening in the delivery room (24%).
- 5 patients were diagnosed between 24H and 48H (24%).
- 2 patients were diagnosed between 48H and 72H (10%).
- 1 patient was diagnosed after 72H (4%).
- The mean diagnostic delay is 17H.
- The extreme delays vary between 0H and 96H. (Figure 7)

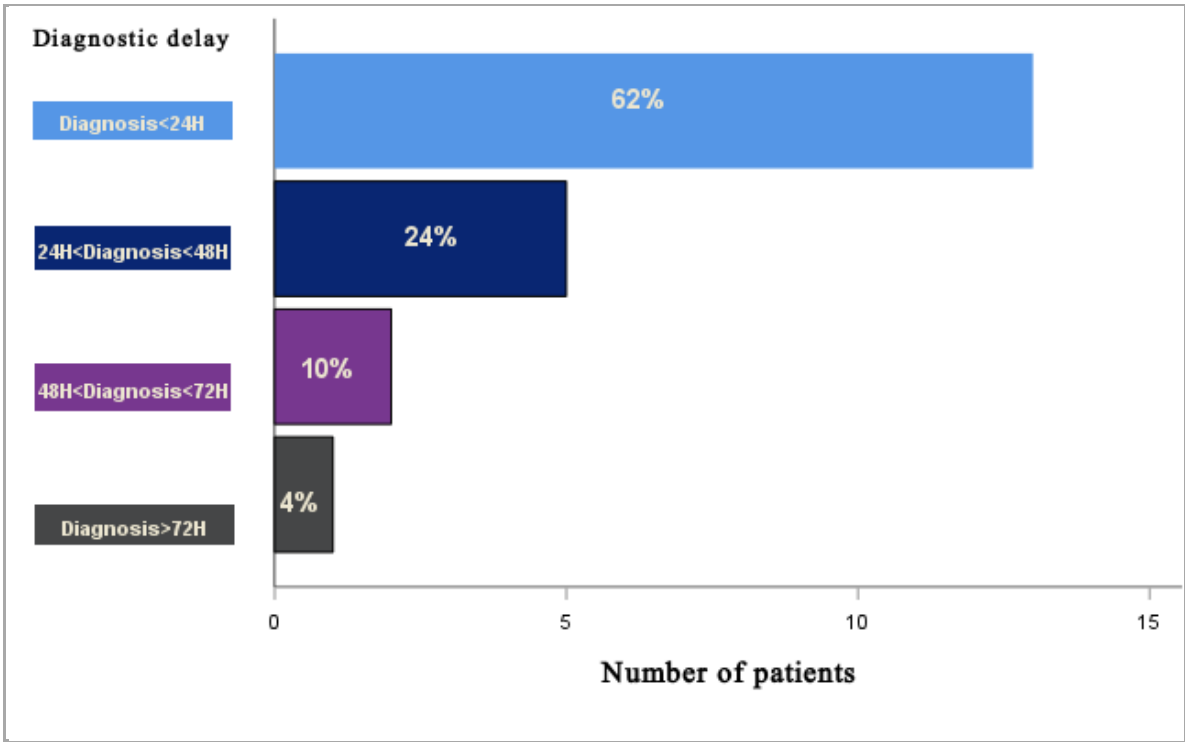


Figure 7 : Cases distribution by diagnostic delay

2. Diagnostic means :

The nasogastric tube insertion test was performed systematically at birth in 5 patients. This allowed a diagnosis at H0 of life in 24% of cases.

The remaining 16 patients were diagnosed after the onset of the clinical signs followed by a positive nasogastric tube insertion test (76%).

The diagnosis of esophageal atresia prenatally was suspected by the presence of polyhydramnios on obstetric ultrasound in 8 cases (38%). (Figure 8)

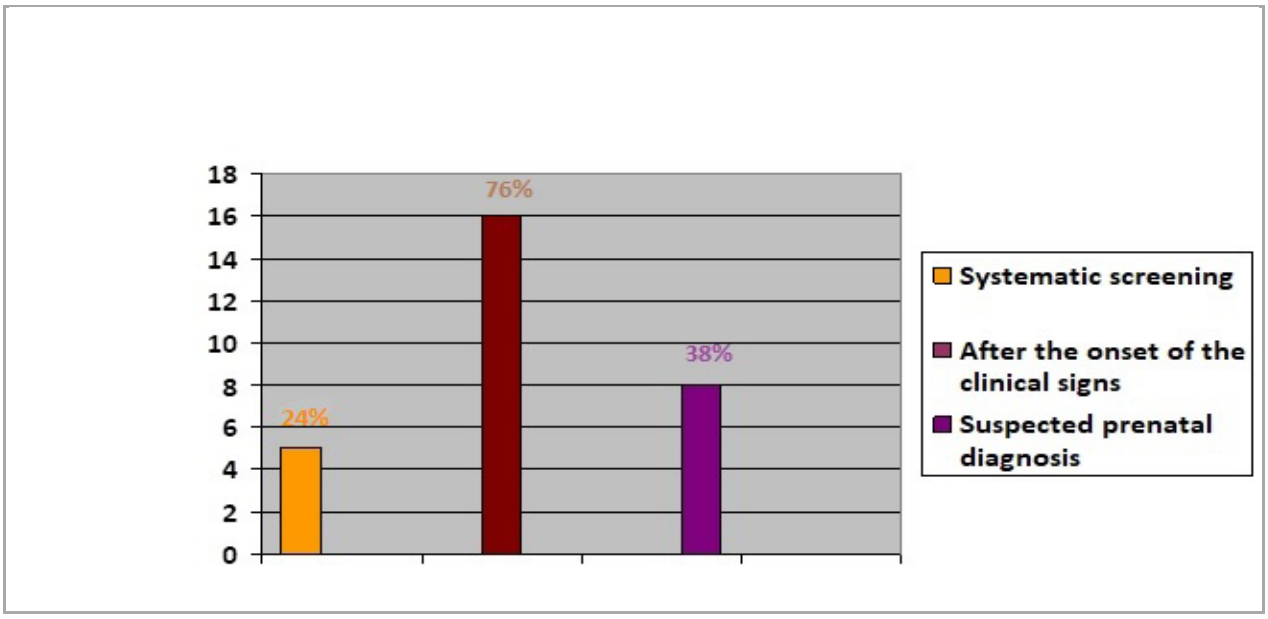


Figure 8 : Patients distribution by diagnostic means

3. Clinical signs :

The most common clinical signs were hypersialorrhea and respiratory distress. Those two signs were observed in 62% of cases. The other clinical signs are summarized in Table V.

An oral feeding attempt was noted in 11 cases (52%).

Table V : Noticed clinical signs

Clinical sign	Number of cases	Percentage
Hypersialorrhea	13	62%
Respiratory distress	13	62%
Cyanosis	6	28%
Episodes of coughing/choking	4	19%
Vomiting	3	14%
Dehydration	1	5%

III. Radiological data :

A frontal chest and abominal X-ray after the placement of a nasogastric tube was systematically performed in all patients. And it showed:

- Coiling of the NG tube in the upper esophageal pouch at a level between the 7th cervical vertebra and the 3rd thoracic vertebra in all our patients X-rays.
- Absence of gastrointestinal air in 13 X-rays (62%), while the presence of air in the gastrointestinal tract was noticed in 8 X-rays (38%).
- The pulmonary status was assessed on all X-rays. A normal pulmonary status was found in 13 cases (62%), and a radiological aspect in favour of a pneumopathy was identified in 8 cases (38%), complicated by pulmonary atelectasis in 3 cases. (Figure 8)
- An aspect of vertebral malformation was noted in 1 case.
- Right aortic arch (RAA) was found in 1 case.
- The gap length between the pouches was measured intraoperatively.

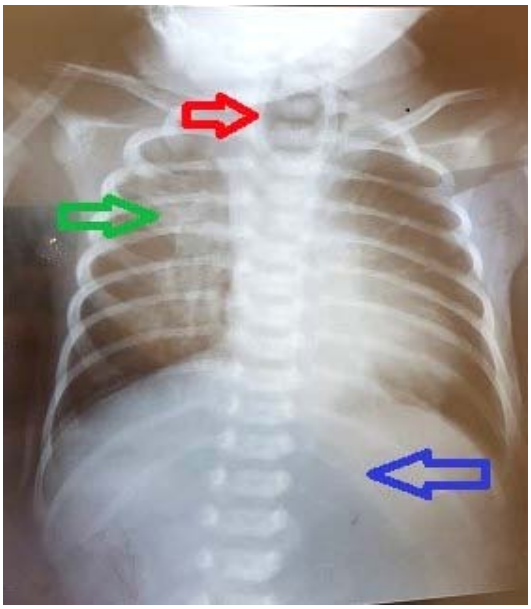


Figure 8 : A frontal chest and abdominal X-ray showing the NG tube coiled in the upper pouch at T3 level (red arrow) with the absence ofair in gastrointestinal tract (blue arrow). We notice the presence of interstitial syndrome in the right middle and superior lobe in favor of a pneumopathy (green arrow).

IV. EA anatomical type :

A pure EA with no fistula (Gross type A) was found in 13 patients (62%). EA with distal tracheoesophageal fistula (Gross type C) was identified in 8 cases (38%). (Figure 9)

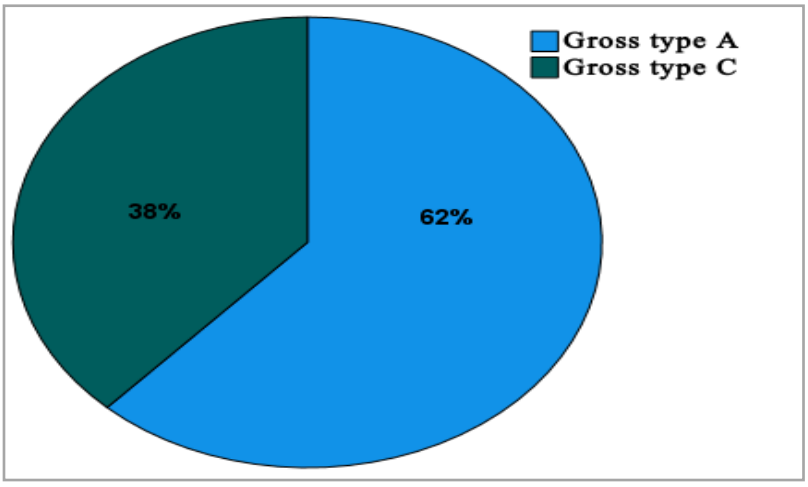


Figure 9 : Cases distribution by EA anatomical type

V. Malformations assessment :

In order to screen various associated malformations, a malformative assessment was performed systematically for all patients. This assessment consisted of: a complete physical examination, a chest and abdominal X-ray, a cardiac ultrasound and an abdominopelvic ultrasound.

2 patients underwent a transfontanellar ultrasound, one because of the presence of a macrocrania found on physical examination and the other because of seizures.

1 patient benefited from a spine X-ray following the suspicion of a vertebral malformation on the chest X-ray.

In our series, 8 patients had associated congenital malformations (38%).

The identified malformations were : cardiovascular, gastrointestinal, genital-urinary, musculoskeletal. (Table VI)

1. Cardiovascular malformations :

In our series, the most common associated malformations occur in the cardiovascular system. Cardiovascular abnormalities were found in 4 patients (19%) :

- Patent ductus arteriosus (PDA) was identified in 2 patients.
- Ventricular septal defect (VSD) was found in 2 patients.
- Atrial septal defect (ASD) was identified in 1 case.
- Coarctation of the aorta (CoA) was found in 1 case.
- Right aortic arch (RAA) was found in 1 case.

2. Genitourinary malformations :

Genital-urinary malformations were identified in 3 cases (14%) :

- Hypospadias was found in 2 cases.
- Unilateral cryptorchidism was identified in 1 case.

3. Gastrointestinal malformations :

Congenital anomalies of the gastrointestinal tract were identified in 2 cases (10%) :

- Duodenal atresia was found in 2 cases.
- Annular pancreas was found in 1 case.

4. Musculoskeletal malformations :

Musculoskeletal malformations were noted in 4 patients (19%) :

- Congenital hip dislocation (CHD) was found in 2 patients.
- Lumbar kyphosis was found in 1 patient.
- Congenital talipes equinovarus (CTE) was identified in 1 patient.
- Radial longitudinal deficiency (RLD) was found in 1 patient.

5. Other malformations :

Triventricular hydrocephalus was found in 1 case, and an ear deformitie (microtia) was found in 1 patient.

6. Malformative association

Among the 8 patients with associated malformations, 3 patients had 2 or more malformations. VACTERL syndrome was found in 1 patient (5%).

Table VI : Associated malformations

Malformations	Number of cases (N)
Cardiovascular (N=4)	
PDA	2
VSD	2
ASD	1
CoA	1
RAA	1
Genital-urinary (N=3)	
Hypospadias	2
Unilateral cryptorchidism	1
Gastroinstestinal (N=2)	
Duodenal atresia	2
Annular pancreas	1
Musculoskeletal (N=4)	
CHD	2
Lumbar kyphosis	1
CTE	1
RLD	1
Other (N=2)	
Triventricular hydrocephalus	1
Microtia	1
Association	
VACTERL syndrome	1

VI. Waterston prognostic classification :

The Waterston classification has been used to stratify all of our patients into 3 prognostic categories (Figure 10) :

- 10 patients were classified in category A (48%).
- 8 patients were classified in category B (38%).
- 3 patients were classified in category C (14%).

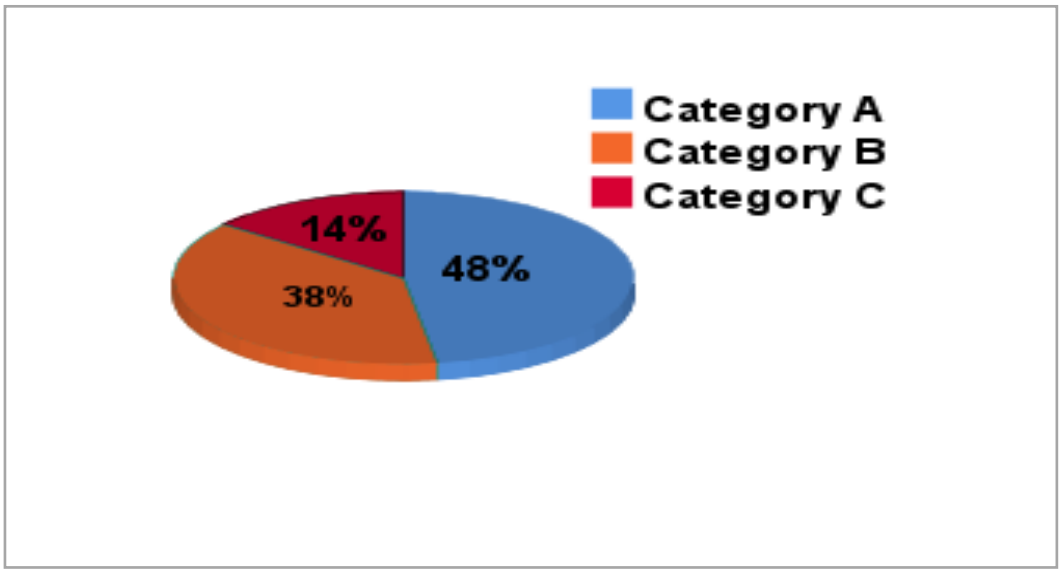


Figure 10 : Patients repartition according to Waterston classification

VII. Therapeutic management :

1. Preparation for surgery :

1.1. Preoperative length of stay in pediatric ICU :

90% of our patients were hospitalized in pediatric ICU prior to surgery.

The average stay length in ICU was 4 days (1 day-16 days).

1.2. Preoperative management :

The preoperative care was as follows :

- Continuous suction after the placement of a nasogastric tube in the upper esophageal pouch to prevent aspiration.
- Placing the patient in a semi-sitting position (45 degrees).
- Cardiorespiratory monitoring.
- Nasal oxygen therapy depending on the oxygen saturation. Patients with sever respiratory distress were intubated.

- Insertion of a peripheral venous catheter.
- Preoperative blood tests consisting of : CBC, CRP, Blood urea nitrogen (BUN), Creatinine, hemostasis test, serum electrolyte test, transaminases, blood glucose level, ABO rhesus typing.
- Administration of vitamine K to prevent the hemorragic disease of the newborn.
- Fluid and electrolytes replacement.
- PPI based therapy was prescribed systematically.
- Antibiotic therapy was administrated when the diagnostic delay exceeded 24 hours with several feeding attempts or signs of infection (signs of sepsis, elevated CRP, leukopenia, thrombocytopenia …). Antibiotic therapy was also prescribed for patients patients whose mothers were diagnosed with PROM.
- Hypothermia prevention for premature newborns.

During their stay in ICU :

- 2 patients had generalized onset seizures secondary to perinatal asphyxia and were treated with phenobarbital.
- 2 patients had neonatal jaundice and were treated with phototherapy.
- 1 patient had functional renal failure secondary to dehydration.
- 1 patient was intubated due a severe respiratory distress.

2. Surgical management :

All patients included in our series were operated.

2.1. Surgical approach :

A systematic midline laparotomy was performed in 13 patients with type A EA for gastrostomy tube placement, and a duodenostomy in 1 patient with duodenal atresia.

For the 8 patients with type C EA, a modified posterior thoracotomy was performed in 7 patients and a left dorsolateral thoracotomy in 1 patient with right aortic arch for the repair of tracheoesophageal fistula and the measurement of the gap length between the proximal and the distal pouch. Completed by a midline laparotomy for gastrostomy.

A lateral cervical approach was performed in 2 patients for establishment of an esophagostomy.

2.2. Surgical exploration :

Surgical exploration has shown :

- Presence of long gap between the proximal and the distal pouch in all patients.
- An annular pancreas in 1 case.
- A right aortic arch in 1 case.
- Hypoplasia of the lower segment in 1 case.
- Duodenal atresia in 1 case.

2.3. Surgical management :

Our therapeutic option was a staged repair of LGEA. Therefore our surgical management was done in 2 stages.

a. First stage :

- Patients with type A EA : underwent systematically a gastrostomy tube placement through a midline laparotomy, 2 patients had undergone, in addition to the gastrostomy, an esophagostomy through a lateral cervical insicion.
- Patients with type C EA : underwent a repair of TEF through a thoracotomy. And when the primary anastomosis was impossible due to the long gap between the pouches, the surgical operation was completed by gastrostomy tube placement through a midline laparotomy. Among these patients, 1 had undergone esophageal elongation by external axial traction (Foker process).

b. Second stage :

8 patients underwent later a definitive repair of LGEA through thoracotomy (39%) :

- Delayed primary anastomosis was performed in 3 cases in which the full continuity of native esophageal tissue was achieved (14%), all of them had a type C EA.
- Colon interposition conbined with pyloroplasty was performed in 2 cases (10%), one had a type A EA and the other a type C EA.
- Gastric transposition associated with pyloroplasty was carried out in 2 cases (10%), one had a type A EA and the other a type C EA.
- Reverse gastric tube was performed in 1 case with type A EA (5%). (Figure 11)

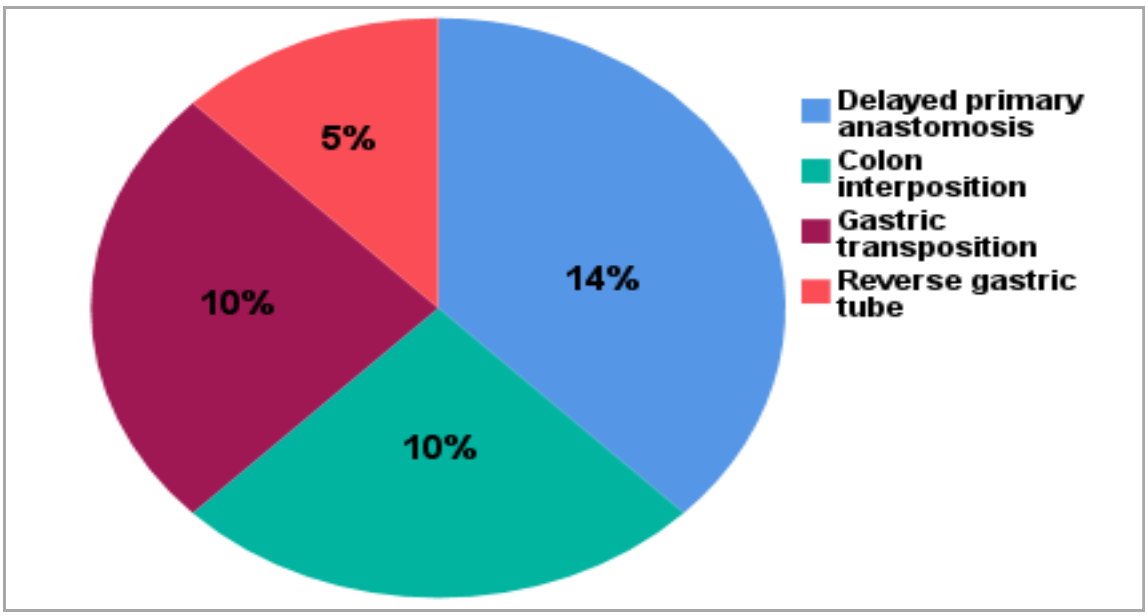


Figure 11 : Surgical options for the definitive repair of the LGEA

VIII. Evolution :

1. Postoperative care :

All patients were routinely admitted to the pediatric ICU after undergoing surgery. (Figure 12)

The average stay length in ICU after establishment of gastrostomy was 6 days (1 day-20 days).

The 8 patients, who underwent a definitive repair of LGEA, had an average stay length in ICU after surgery of 12 days (2 days-32 days).

During the stay in ICU, the following measures were undertaken:

- Placing patients in a semi-sitting position (45 degrees).
- Placement of an oropharyngeal tube for continuous suction.
- Cardiorespiratory monitoring.
- Fluid and electrolytes replacement.
- Multimodal analgesia combining paracetamol and morphine.
- Antibiotic prophylaxis, PPI and antiemetic were systematically prescribed.
- Daily respiratory physiotherapy.
- Enteral feeding was initiated through a nasogastric tube after an average of 3 days post gastrostomy.
- Patients, who underwent definitive repair of LGEA, were extubated and fed orally after a swallow esophagogram showing the integrity of the anastomosis, on average 7 days after the surgery.
- Completion of the malformations assessment.

Figure 12 [2] :Picture of a patient, one day after undergoing the definitive repair of LGEA type C with cervical flexion, in spontaneous breathing with inspiratory pressure support (white arrow shows the extrapleural chest tube)

2. Postoperative complications :

7 patients had an uneventful postoperative course (33%), while 14 patients presented postoperative complications listed in Table VII. (Figure 12)

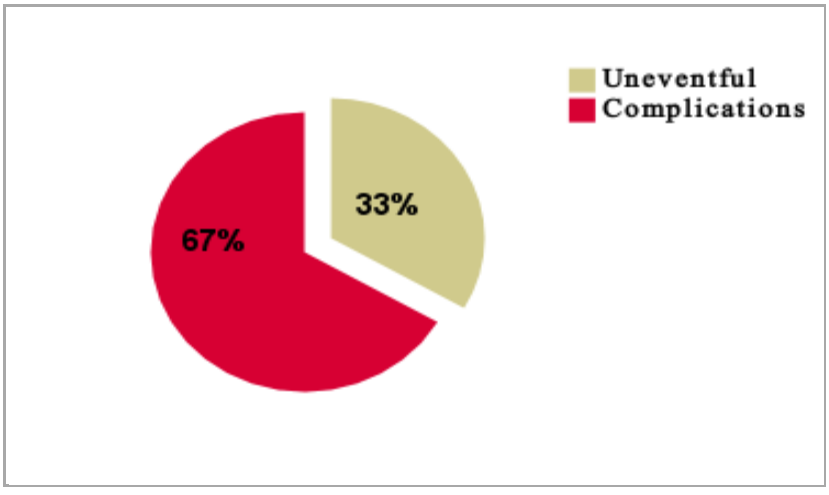


Figure 13 : Prevalence of postoperative complications

Table VII :Postoperative complications

Postoperative complications	Number of patients	Percentage
Anastomotic leakage	3	14%
Fistula recurrence	1	5%
Wound infection	2	10%
Sepsis	4	19%
Necrotic lesion of the foot	1	5%
Nosocomial pneumonia	5	24%
Respiratory distress	4	19%
Apnea	1	5%
Pneumothorax	2	10%
Pneumomediastinum	1	5%
Acute renal failure	1	5%
Hypoxic cardiac arrest	1	5%

For the 3 cases of anatomotic leakage, 2 patients were put under continuous suction and antibiotic therapy. While the 3rd patient underwent another intervention to re-establish the hermicity of the gastrostomy and to perform a esophagostomy.

The case of fistula recurrence was reoperated after receiving a continous suction to prevent reflux and aspiration pneumonia.

IX. Mortality :

1. Mortality rate :

In our series the number of deceased patients was 9 patients. The mortality rate is 43%. (Figure 14)

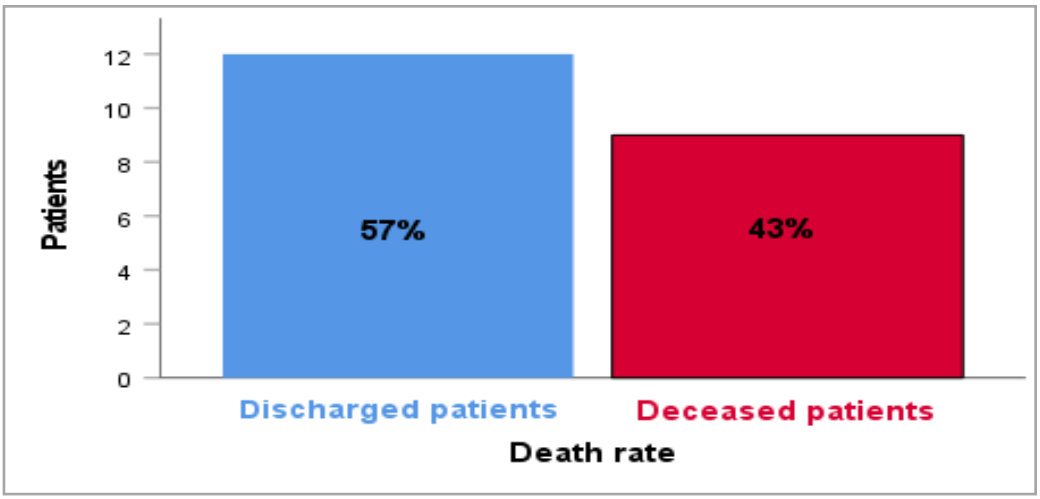


Figure 14 : Mortality rate

The most common cause of death is nosocomial pneumonia complicated by septic shock.

2. Survival time :

Among the 9 deceased patients, 5 patients passed away after undergoing the initial gastrostomy (56%). While the other 4 patients passed away after undergoing the definitive repair of LGEA (44%).

The average survival time after the initial gastrostomy was 78 days (4 days-184 days).

The average survival time after the definitive repair of LGEA was 25 days (1day -62 days).

3. Waterston classification :

4 deceased patients were classified in category B (44%), 3 in category A (33%) and 2 in category C (23%). (Table VIII)

Table VIII : Deceased distribution by Waterson classification

Waterson categories	Number of patients	Percentage
Category A	3	33%
Category B	4	44%
Category C	2	23%

4. Diagnostic delay :

- 4 of the 9 deceased patients were diagnosed within the first 24 hours (44%).
- 3 patients were diagnosed after a delay between 24 and 48 hours (34%).
- 1 patient was diagnosed after a delay between 48 and 72 hours (11%).
- 1 patient was diagnosed after a delay exceeding 72 hours (11%). (Table IX)

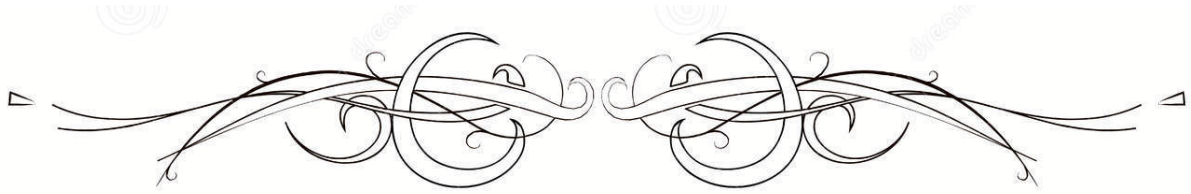
Table IX : Deceased patients distribution by diagnostic delay

Diagnostic delay	Number of patients	Percentage
Dg<24H	4	44%
24H<Dg<48H	3	34%
48H<Dg<72H	1	11%
Dg>72H	1	11%

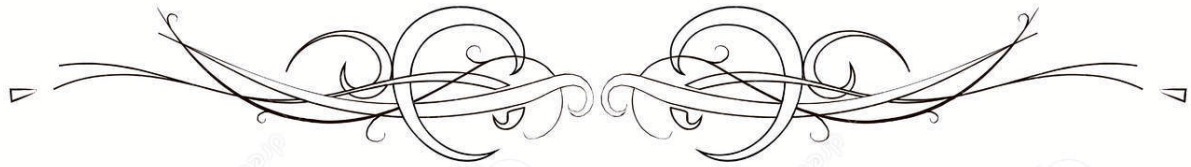
5. Postoperative complications :

Among the 9 patients who passed away :

- 4 patients had a nosocomial pneumonia complicated by sepsis.
- 3 patients had an acute respiratory distress.
- 2 patients had an anastomotic leakage.
- 2 patients presented a dehydration.
- 1 patient had pneumothorax.
- 1 patient had pneumomediastinum.
- 1 patient had hypoxic cardiac arrest.



DISCUSSION



I. Definition :

Esophageal atresia (EA) is defined as an interruption in the continuity of the oesophagus with or without fistula to the trachea. It is considered the most common congenital anomaly of the esophagus [3].

The diagnostic criteria for long gap differ among different surgeons. Some define 2 cm as a cut-off point, others classify the gap into short (1 cm), intermediate (2.5–3.0 cm) and long (>3 cm), others define a gap more than 3–3.5 cm as long, while still others recommend an esophageal replacement if the gap exceeds the length of six vertebral bodies. There is also a lack of uniformity in the methods used to measure the gap. In addition, most reports do not indicate whether the gap was measured before or after dissection of the esophageal stumps, or whether or not it was measured under tension [4].

The method of gap measurement chiefly depends on the anatomical type of esophageal atresia, and is usually done under general anesthesia. In patients with type A or B esophageal atresia (as well as in those with a previous failed attempt), the gap between the esophageal ends may be measured either by injecting sufficient radio-opaque contrast into the stomach to allow it to enter the distal esophagus, or by passing a radio-opaque instrument, such as a bougie, Hegar dilator, urethral sound or flexible endoscope, through the gastrostomy site into the distal stump. At the same time, a radio-opaque tube is advanced in the upper pouch. The use of rigid tools allows measurement of the gap both under passive tension (not pushing on the rigid instruments) and with active stretch (pushing on the instruments) and gives a more precise estimate of the true gap and the degree of tension that can be anticipated after anastomosis [4]. (Figure 14)

In patients with type C esophageal atresia, it might not be possible to measure the gap with the method described above, due to the absence of gastrostomy. In such cases, a preoperative estimate of gap length can be made by measuring, under fluoroscopy, the length between the radiopaque tube deeply inserted in the blind upper pouch and the broncoschope's

tip just placed at the opening of lower fistula. Otherwise an intra-operative measurement, both with and without tension on the esophageal ends, is performed after tracheo-esophageal fistula division [4].

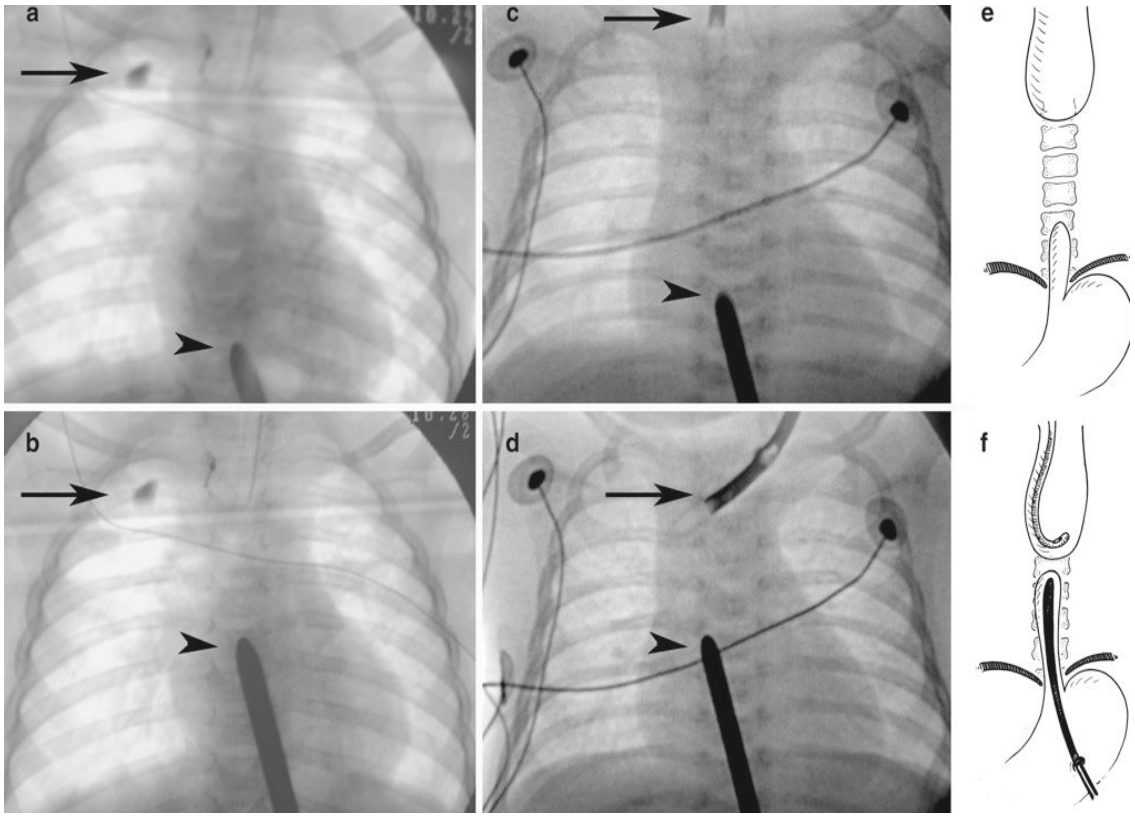


Figure 14[4] : Gap measurement (gapogram) in type A esophageal atresia.

(a) Without tension in a patient with cervical esophagostomy. Arrow: Contrast media at the level of the cervical esophagostomy. Arrowhead: Hegar dilator passed through the gastrostomy.

(b) Gapogram of the same patient as under tension. Arrow: Contrast at the level of the cervical esophagostomy. Arrowhead: Hegar dilator passed through the gastrostomy. Note the reduction of the gap under active tension on the lower esophagus.

(c) Gapogram without tension. Arrow : Tip of the radio-opaque tube inserted in the upper esophageal pouch. Arrowhead:Hegar dilator passed through the gastrostomy.

(d) Gapogram of thesame patient as (c) under tension on both esophageal pouches. Arrow : Tip of the radiopaque tube inserted in the upper esophageal pouch. Arrowhead: Hegar dilator passed through the gastrostomy. Note the reduction of the gap under active tension.

(e, f) Diagram showing a type A esophageal atresia without tension on the pouches (e) and after a Hegar dilator is passed through the gastrostomy and active tension is applied on both esophageal pouches.

II. Epidemiology :

1. Frequency :

Long-gap esophageal atresia (LGEA) is a rare malformation that occurs approximately 10% of children born with esophageal atresia (EA) [5] which has a prevalence of 1 in every 2500-4500 live births [6].

The French national EA registry was created in 2008 by the reference center for congenital abnormalities of the esophagus. It differs from other national registries because it is specific for EA. Its goal is to evaluate the prevalence of this congenital malformation in the children born alive in France and their evolution during infancy. It also collects prenatal data, neonatal treatment and early outcome, as well as follow-up for the first year of life. It is based on a national network of all 38 French centers performing neonatal surgery. The prevalence of EA in 2008 and 2009 in France was 1.97 per 10 000 live births. The percentage of pure EA, which is a variant of LGEA, was 10% of all recorded EA cases [7].

In the series of Jensen et al. [1], in which a retrospective review of 218 EA patients from 1994 to 2016 was undertaken, 37 patients had a LGEA, representing a percentage of 17%.

In Morocco, the prevalence of LGEA is poorly estimated due to the absence of a national registry of congenital malformations and also due to the fact that our study is the first study in the country to focus solely on LGEA. (Table X)

Table X : Frequency of LGEA in some Moroccan series

Series	Number of cases/ Study duration	Frequency per year
Our series	21 cases/7 years	3 cases/year
Series of Benkirane Marrakech 2020 [2]	6 Cases/2 years	3 cases/year
Series of Mahmoudi Oujda 2017 [8]	32 cases/6 years	5,3 cases/year
Series of Loughlimi Marrakech 2012 [9]	6 cases/2 years	3 cases/year

2. Gestational age :

Prematurity has a significant influence on outcome of newborns with LGEA. Preterm newborns are at high risk of morbidity and mortality [14].

The prematurity rate in our series was 19%, which is a low rate compared to the series of Jensen et al. (48,6%) [1] and the series of Seguier-Lipszyc et al. (50%) [15]. The highest prematurity rate was recorded in the series of Lee et al. (57%) [16]. (Table XII)

This high prematurity rate associated with EA is most likely related to polyhdramnios, which is often found in EA [17].

Table XII :Comparison of prematurity rate with other series

Series	Number of patients	Prematurity rate
Our series	21	19%
Series of Jensen et al.[1]	37	48,6%
Series of Seguier-Lipszyc et al.[15]	10	50%
Series of Lee et al. [16]	30	57%

3. Birth weight :

The average birth weight in our series is 2790 grams, which is higher than all the series listed in the Table XIII.

Table XIII : Comparison of our series mean birth weight with other series

Series	Number of patients	Mean birth weight
Our series	21	2790 g
Series of Gallo et al. [18]	24	2017 g
Series of Hunter et al.[19]	28	2217 g
Series of Holland et al. [20]	33	2327 g

In infants with EA, birth weight has consistently been reported as a significant determinant of overall survival and it has been included as a criterion in several prognostic classifications. In 1962, Waterston et al. from Great Ormond Street reported the outcomes of 218

infants born with EA and proposed a prognostic classification based on birth weight, associated anomalies and pneumonia [21]. In that study, the lowest rates of survival were in infants with birth weights less than 1800 g. In 1994, Spitz et al. updated the prognostic classification after reviewing 372 infants with EA, with the lowest survival rates in infants with major cardiac anomalies and birth weights less than 1500 g [22]. More recently, other groups reviewing smaller series of EA patients have proposed modifications of these classifications with birth weight becoming a less significant determinant for survival compared to the Waterston and Spitz series [23], [24]. Poenaru et al. reported that birth weight was not found to independently influence mortality in their series of 96 patients and proposed a novel "Montreal classification", based on two other prognostic factors, pre-operative ventilator dependence and associated anomalies [25]. Similarly, Choudhury et al. reported that associated cardiac and chromosomal anomalies were more significant causes of death than birth weight [26]. Conversely, a more recent Japanese study analyzing the outcome of EA infants treated over 25 years reported a modification of the Spitz classification where birth weight of less than 2000 g and major cardiac anomalies were still considered crucial factors for survival [27]. However, these studies propose new classifications based on relatively small cohorts of patients accrued over a very long period of review during which many facets of neonatal care have changed or improved [28].

4. Maternal medical history :

The etiology of EA is complex and heterogeneous. A nationwide, population-based, case-control study was nested in a cohort of children born in Sweden from 1982 to 2004 to establish a causal link between maternal tobacco smocking, maternal obesity and the occurrence of EA.Among 2 305 858 newborn children constituting the study cohort, 722 cases of EA and 3610 controls were included. For women smoking 10 cigarettes or more daily, the adjusted OR (Odds ratios) was 0.88 compared to nonsmokers. For obese women (body mass index, >30), OR was

0.99 compared to lean women (body mass index<20). This study provides evidence refuting the hypotheses of an increased risk of EA among children of obese or/and smoker women [29].

Another population-based, matched case-control study, nested within a cohort of neonates born in Sweden in January 1982 through December 2007, was undertaken to demonstrate the responsibility of maternal diabetes in the EA and associated malformations development. Among 2 625 436 newborn infants in the study cohort, there were 780 cases of esophageal atresia, and 7800 infants were matched and randomly selected as controls. The adjusted risk of esophageal atresia was 70 % higher among infants of women with diabetes than among women without diabetes.However, the study could not show a causal link between diabetes and malformations associated with EA. In addition, it could not determine whether the risk of esophageal atresia was higher in gestational diabetes than in pre-existing diabetes [30].

5. Parity and maternal age :

In our series the primiparity rate was 52%. The average maternal age could not be evaluated due to the lack of information in our patients files.

A study examined the prevalence of EA and the relationship between EA and demographic factors in the Russian Federation. Data were obtained from a population-based congenital malformations registry across 14 years (2000–2013) in 24 regions of the Russian Federation and included cases of EA among live births and stillbirth. It was shown, at the 5% level, that there was no influence on the risk of EA when maternal age was under 19 years old. However, the risk was higher for mothers older than 35 years and for the first gravidity [31].

A Swedish nationwide case-control study [32] has shown similar results. With regard to maternal parity, it turned out that 32%, 36%, and 21% decrease in the risk of delivering a child with EA among women giving birth to their second, third, or fourth or more child, respectively, compared to primiparous. Regarding maternal age, women giving birth between the ages of 35 and 40 years had an adjusted 2-fold increase in risk of delivering a child with EA compared to

mothers younger than 20 years, and the corresponding risk was increased 3-fold among mothers older than 40 years.

On the other hand, the series of Bianca [33] showed that the risk of occurrence of EA increases with the number of parity.

While some studies have shown no association between maternal age, parity and EA development [34].

III. Anatomical classification :

Several anatomical classifications of EA have been established based on the presence or not of a tracheoesophageal fistula and on the location of the fistula.

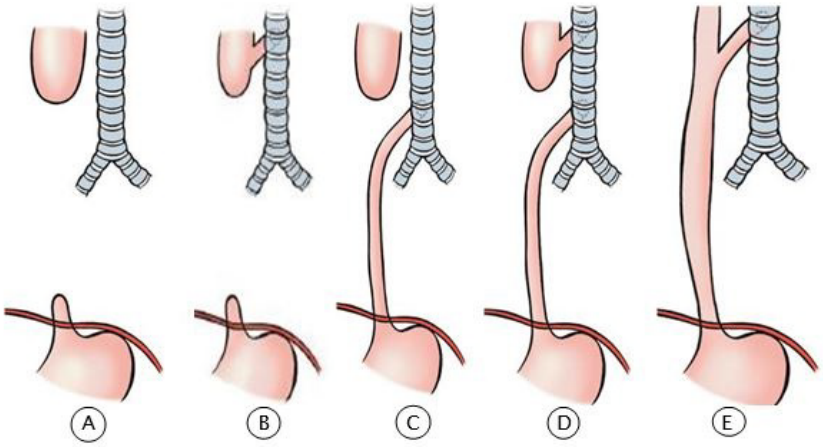
In 1929, Vogt[35] first proposed an anatomical classification of EA and TEF based on radiological and postmortem findings. Ladd[36] put forth his own classification in 1945 and Gross [37] revised this schema in 1953.

In 1976, Kluth [38] estbalished the most detailed classification which incorporates all described anatomical variants of EA and TEF.

The different anatomical classifications with their subtypes are summarised in Table XIV and Figure 15.

Table XIV : EA anatomical classifications subtypes

Vogt	Ladd	Gross	Description	Frequency
Type 1	-	-	Esophageal agenesis	N/A
Type 2	I	Type A	EA without TEF	7%
Type 3A	II	Type B	EA with proximal TEF	2-3%
Type 3B	III,IV	Type C	EA with distal TEF	85%
Type 3C	V	Type D	EA with both proximal and distal TEFs	<1%
Type 4	-	Type E	TEF without EA, H-type	4%



A : EA without TEF
B : EA with proximal TEF
C : EA with distal TEF
D : EA with proximal and distal TEFs
E : TEF without EA

Figure 15 [39] : Classification of EA/TEF

The most frequent anatomical type in our series is Gross type A with a percentage of 62%, which is consistent with the results of all series in Table XV.

Table XV : Anatomical types in different series according to Gross system

Series	Type A	Type B	Type C	Type D	Type E
Our series	62%	-	38%	-	-
Series of Jensen [1]	62%	16%	22%	-	-
Series of Stadil et al. [40]	78,9%	21,1%	-	-	-
Series of Bagolan et al. [41]	46%	10%	44%	-	-

IV. Diagnosis :

1. Prenatal diagnosis :

In most patients, EA is diagnosed after birth. The prenatal detection of EA range between 9,2% and 40%. In some studies it was reported that the rate of prenatal diagnosis of LGEA was much higher than that of non-LGEA [42].

The advantage of antenatal diagnosis is to schedule the delivery in a hospital with a neonatal intensive care unit and a pediatric surgical team capable of operating on this malformation and thus avoid delays in management and improve the prognosis. However, some authors has shown that the prognosis of LGEA does not appear to be influenced by any prenatal diagnosis [43].

The diagnosis of LGEA was suspected antenatally by the presence of polyhdramnios in 38% of our cases, which is lower than the rate in the series of Garabedian et al. [43] and the series of Spaggiari et al. [44], and significantly higher than that in the series of Erikci et al. [42]. (Table XVI)

Table XVI : LGEA prenatal detection rate in several series

Series	Number of patients	Prenatal detection rate
Our series	21	38%
Series of Erikci et al. [42]	18	22,2%
Series of Garabedian et al.[43]	88	85,5%
Series of Spaggiari et al. [44]	14	85,7%

1.1. Obstetrical ultrasound :

Prenatal diagnosis is difficult due to the fact that normal esophagus is usually not seen in routine ultrasound examination because its collapsed lumen and tissue textures are similar to those of other organs. However, Malinger et al [45] reported visualization of normal esophagus using a high-resolution linear transducer in a prospective study concerning 60 consecutive fetuses between 19 and 25 weeks gestation, with complete visualization of the esophagus in 86.7% of cases and partial visualization in 96,7%. (Figure 16)

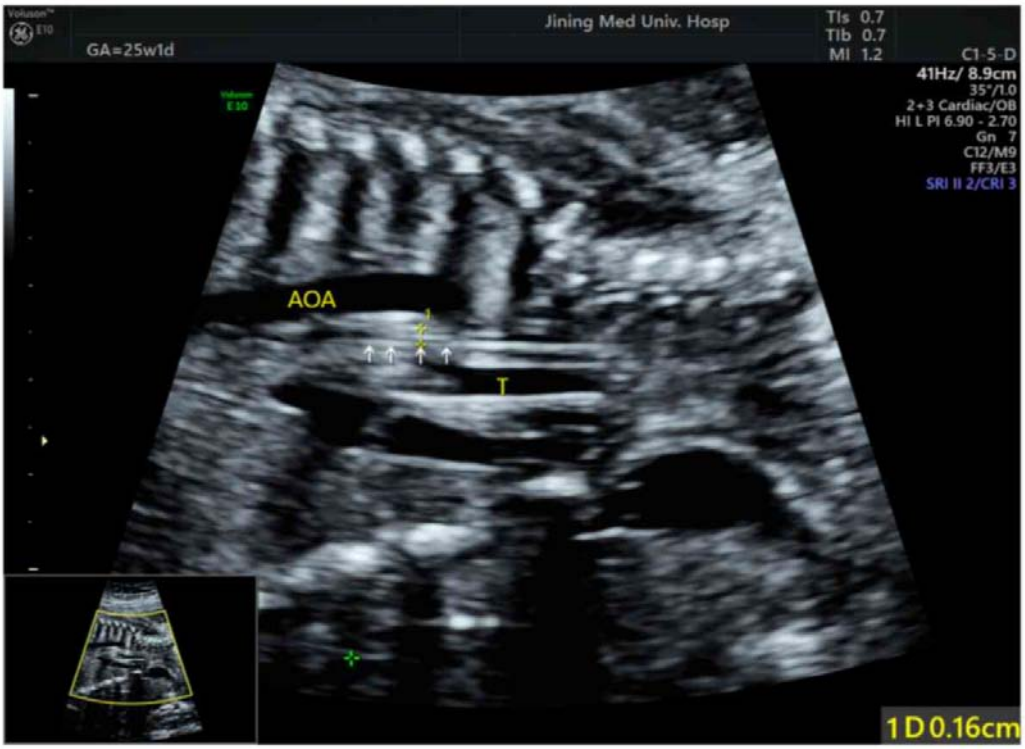


Figure 16 [46] : Normal unfilled esophagus of a fetus in a prone position. The white arrows indicate the esophagus and calipers 1 shows the diameter of the unfilled esophagus. AOA : aorta; T : trachea.

Diagnosis of EA is usually suspected in the third trimester because of indirect signs: association of polyhydramnios and absent or small stomach bubble. Because polyhydramnios rarely develops before 24 weeks of gestation, the time of diagnosis is often late. Isolated polyhydramnios, and absent or small stomach bubble are poor predictors of EA. Most EAs have a TEF (90%) with possible visualization of the fetal stomach because of amniotic fluid passing in the stomach through the fistula. Even with no fistula, the stomach may be visualized because of secretions from the gastric mucosa [47]. Polyhydramnios alone is not a good sign of EA due to the fact that EA is the cause of only 1% of polyhydramnios. Conversely, polyhydramnios is not always present in cases of EA.

The predictive positive value of the combination of these 2 signs is low, between 40% and 56%, with many infants suspected prenatally to have EA with a normal esophagus at birth. Numerous other causes such as neuromuscular deficit, central nervous system anomalies, and

facial anomalies could lead to absent or small stomach bubble and polyhydramnios because of impaired fetal swallowing. The false-positive rate could even be higher in fetuses with multiple anomalies.

In the study by Choudry et al [48], EA was confirmed in only 2 of 25 cases of small or absent stomach bubble (Figure 17) associated with other anomalies (false-positive rate 77.5%: 47% in cases of suspicion of isolated EA and 92% in cases of suspicion of nonisolated EA).

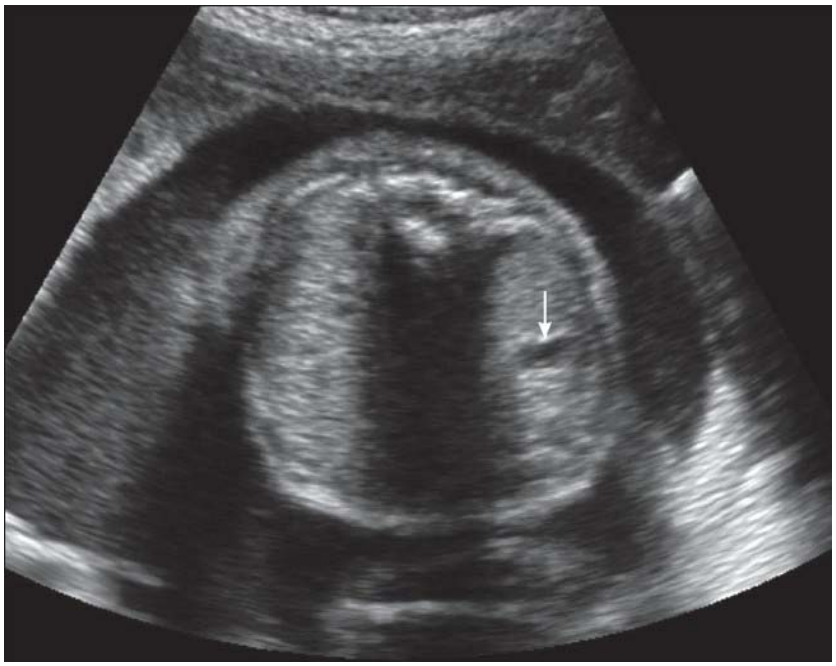


Figure 17 [49] : Ultrasound of a fetus in the transverse plane at the level of abdomen shows the small stomach (white arrow)

The prenatal sonographic diagnosis can be improved by direct visualization of the fluid-filled, blind-ending esophagus during fetal swallowing or upper neck “pouch sign. The pouch may be in the cervical region "neck pouch" (Figure 18) or in the upper mediastinum "mediastinal pouch" (Figure 19) below the clavicles. This differentiation is important with respect to postnatal repair. Pouches with the base situated in the thorax are always associated with a distal tracheoesophageal fistula and primary esophageal repair is possible. In pouches situated higher, with the base in the neck, primary repair was not possible. This is because neck pouches are associated with a longer atretic gap and a higher probability of severe associated anomalies.

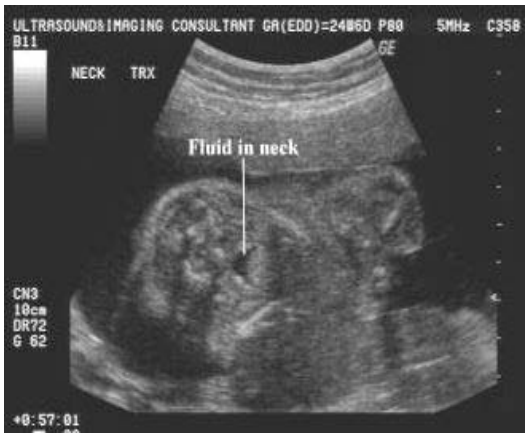


Figure 18 [50] : Neck pouch sign

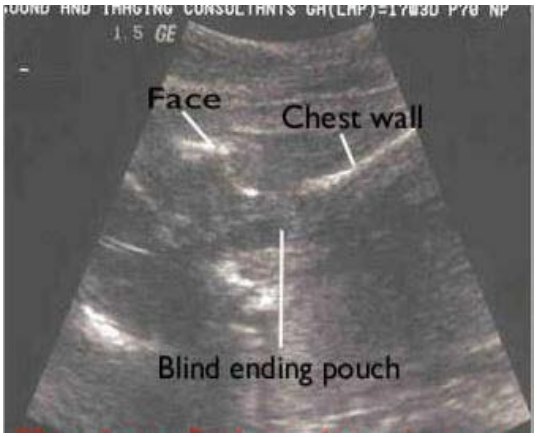


Figure 19 [50] : Mediastinal pouch sign

The pouch sign was first described prenatally in 1983. It has a high specificity for EA because all of the reported cases have postnatally been confirmed to have EA [47].As polyhydramnios, this sign is described most of the time during the third trimester of gestation, which may be because the fetus is not able before to develop sufficient pressure in swallowing to dilate a blind esophagus [50]. However, it has sometimes been observed during the second trimester as early as 23 weeks [50], [51]. To identify the pouch, multiple ultrasonographic sections of the fetal neck and chest region are necessary [51]. Shulman et al [51] developed a systematic approach to 2D ultrasound scanning of the fetal neck and chest in fetuses suspected of having EA (3 image plans of the neck and upper chest: coronal, sagittal, and axial views in the cephalic direction). A low-level pouch (below the clavicle) was better visualized on a sagittal view, whereas the coronal view was optimal for high pouch. Research of the pouch sign implies a longer examination period (20–30 minutes) often performed at repeated short time intervals. Its identification depends on fetal position and mobility, gestational age (rare before 26 weeks), and the presence of fetal swallowing during examination. Consequently, failure to identify a pouch in the fetal neck does not exclude EA [50], [52]. In the study by Brantberg [53], in 21 cases of prenatally diagnosed EA, only 43% had a visible esophageal pouch. Yagel et al [54] reported the use of 3D ultrasound acquisition and postprocessing rendering to visualize the esophageal pouch. Three- and 4D ultrasound acquisition could be useful to decrease time examination and increase sensitivity.

The association of these 3 signs (polyhydramnios, absent or small stomach, and presence of a pouch) (Figure 20), especially if they persist in successive examinations, increases the likelihood of EA. The positive predictive value of this association ranges from 60% to 100% with 80% to 100% sensitivity [45], [51]. Each time EA is suspected because of the association of small stomach and polyhydramnios, a careful examination of fetal neck and chest must be performed on repeated and prolonged scans to improve the EA detection rate [50]–[52].

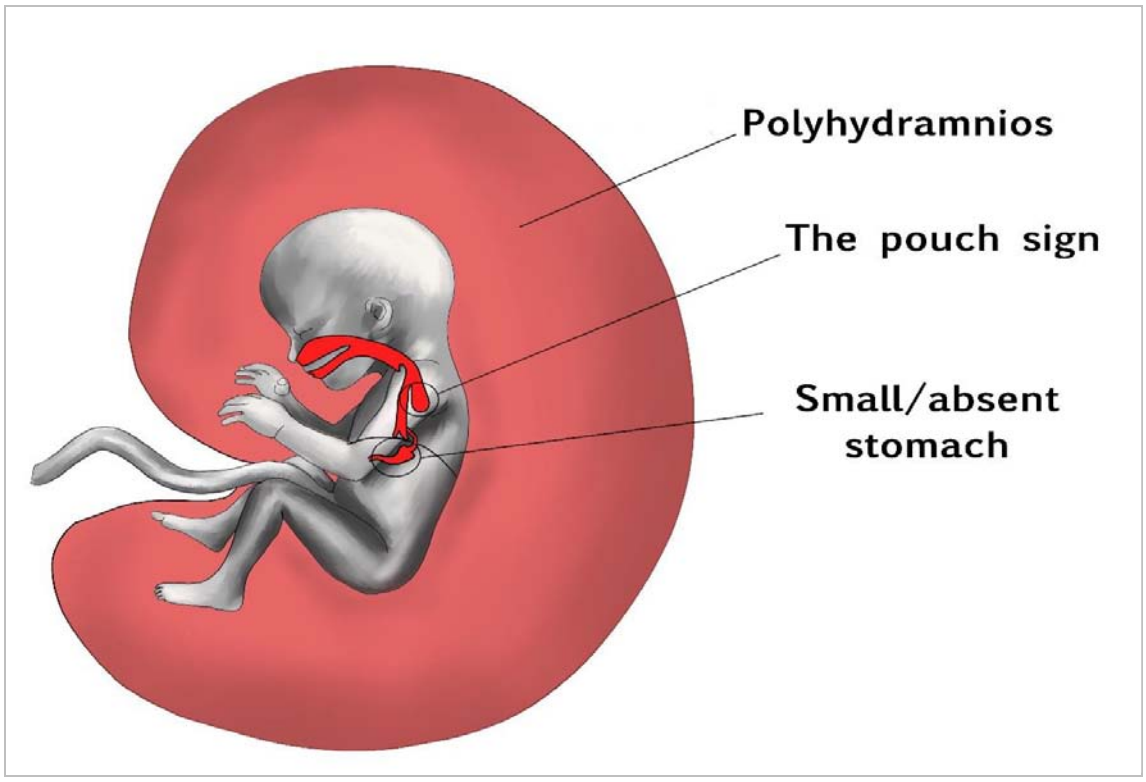


Figure 20 [55] : An illustration of the common prenatal signs of EA

The distended fetal hypopharynx is a novel prenatal sign of EA. It is more sensitive than the pouch sign and at gestational age>28 weeks, has a better predictive accuracy than the pouch sign or any combination of secondary signs according to Tracy et al. [56]. (Figure 21)

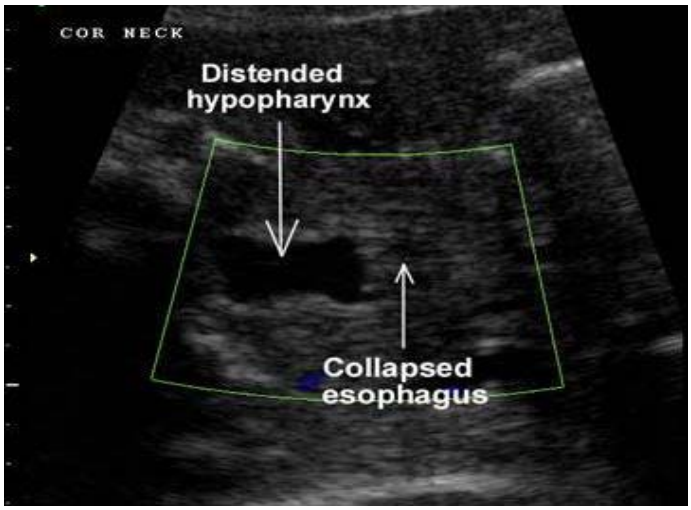


Figure 21[56] : Distended fetal hypopharynx

1.2. Fetal MRI :

MRI allows to overcome the difficulties related to the fetal position and maternal echogenicity. Its objective is to confirm the diagnosis of EA, but also to search for associated anomalies, which were difficult to identify on fetal ultrasound, such as anorectal malformations and the CHARGE syndrome. The location of the midline sagittal plane may be easier in MRI.

For Langer et al. [57] the diagnosis of EA is confirmed in case of non visualization of the intra thoracic part of the esophagus in T2 weighted sequence (100% sensitivity, 80% specificity and 83% positive predictive value in a series of 10 EA ultrasound suspected and 5 confirmed).

However, in the Levine et al. [58] study on 85 MRIs for thoracic abnormalities, the esophagus was not identified in 64% of cases. Therefore, nonvisualization of the entire esophagus as the only diagnostic criterion for EA would lead to many false positives. In the study by C. Garabedian [43], three fetuses had EA: in one case the thoracic portion of the esophagus was visualized, while in the other two cases the esophagus was not.

Other signs have been described on MRI such as dilatation of the hypopharynx, bowed trachea, absence of the stomach bubble or direct visualization of the fistula (one case report) [59].

Visualization of the pouch sign is certainly easier with MRI and functional MRI sequences allow for improved diagnosis by visualizing fetal swallowing movements and highlighting upper pouch dilatation.

However, the use of MRI has some limitations. As in ultrasound examination, fetal swallowing is necessary to visualize the pouch, MRI is not easily available in all of the centers, and the duration of the examination could be a limitation because of the discomfort of the patient in the supine position or excessive fetal movement in cases of polyhydramnios [52].

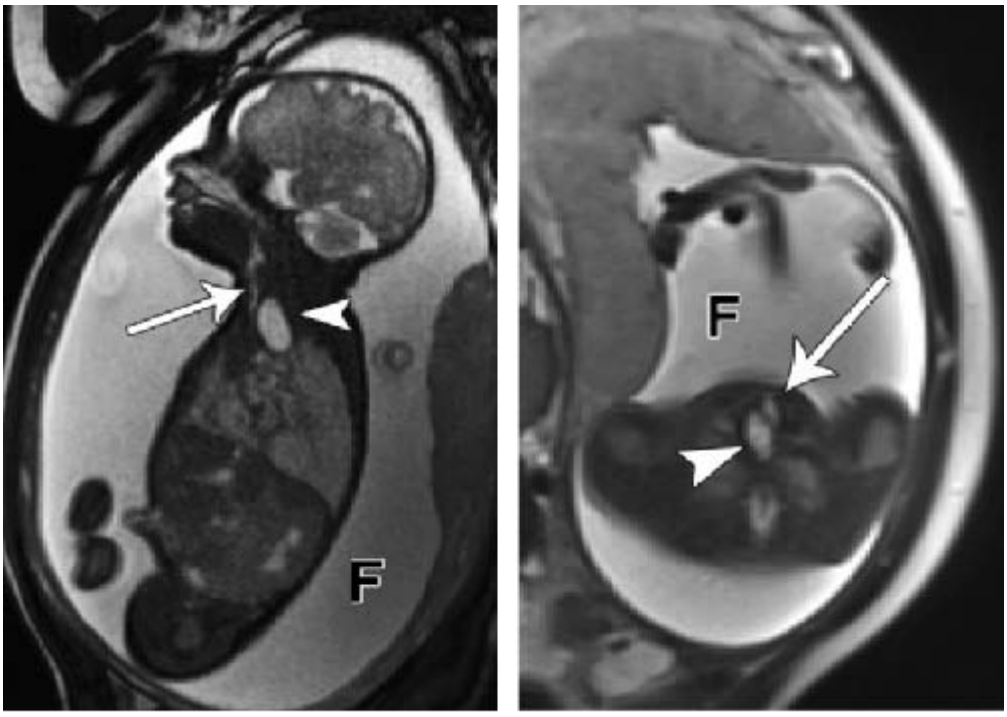


Figure 22 [60] : T2-weighted sagittal sequence through the fetal chest and abdomen (a) and axial T2-weighted sequence (b) show a large esophageal pouch (pouch sign) in the superior thoracic cavity (arrowhead) posterior to the trachea (arrow).

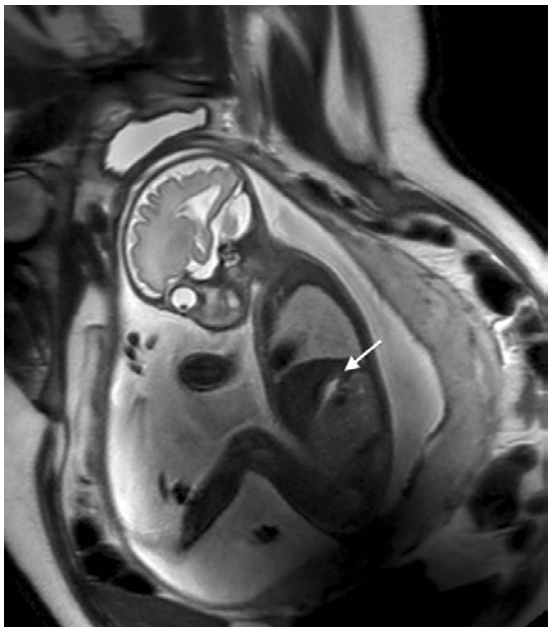


Figure 23[61] : Small size stomach

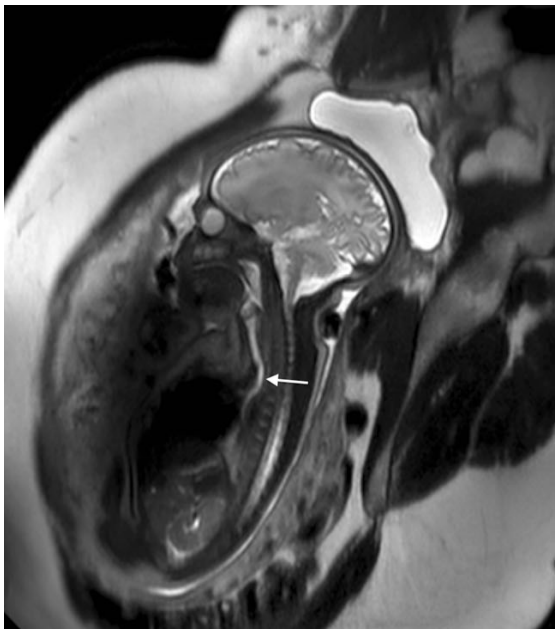


Figure 24[61] : Bowed trachea

1.3. Biochemical markers :

Due to the observed swallowing abnormalities in EA, it is possible to study changes in digestive enzymes in amniotic fluid starting from 18 SA[62].

In Czerkiewicz et al [62] study, 44 fetuses with EA were compared with 88 cases of polyhydramnios and 88 cases of fetuses with normal amniotic fluid. Proteins in the amniotic fluid, alpha fetoprotein (AFP) and digestive enzymes including gamma-glutamyl transpeptidase (GGTP) were assayed for each fetus. An EA index was developed (AFP X GGTP). Using a cutoff of 3 for the EA index, 98% sensitivity and 100% specificity were observed for amniotic fluid prenatal diagnosis of EA, whatever the anatomical type.

These results are extremely interesting as amniocentesis is often performed in cases of suspected EA to verify the presence of chromosomal abnormalities. Amniotic fluid analysis can also be performed during amnioreduction in the context of threatened preterm labor.

2. Postnatal diagnosis :

EA screening must be systematic in the examination of all newborns in the delivery room. An early diagnosis before the outset of any clinical sign, and a fortiori before any oral feeding attempt, allows the necessary measures to be taken to prevent any potential pulmonary complications.

To confirm the esophageal continuity, a NG tube is inserted through the nose or the mouth. Diagnosis is highly suspected by the failure of passage of the NG tube. A characteristic resistance is felt at the blind-ending upper esophageal pouch, and the tube cannot be introduced into the stomach. However, this maneuver is subject to diagnostic errors:

- If the tube is too thin or soft, it may roll up into the pharynx or in the upper pouch, giving the false impression that it has been pushed into the stomach without perceiving the characteristic resistance.
- The passage of the NG tube through the TEF makes a false impression of an intact esophagus [22].
- If the tube is too rigid or too sharp, it may fail to pass when there is actually no EA [63].

A plain chest and abdominal X-ray confirms the diagnosis and demonstrates the NG tube coiled in the upper pouch. An associated TEF is confirmed by the presence of gas-filled intestinal loops below the diaphragm (Figure 25). In isolated or pure EA, a featureless gasless abdominal X-ray is observed (Figure 26).

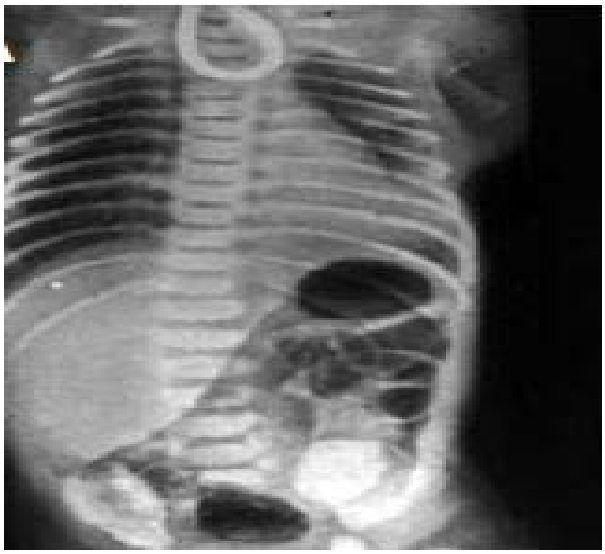


Figure 25 [64] : Coiling of the NG in the upper pouch and presence of air in the stomach indicating the presence of a distal TEF.

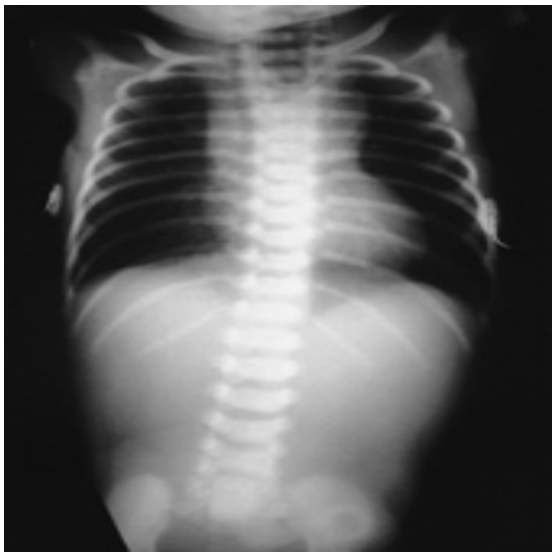


Figure 26 [14] : The NG tube is coiled in the upper pouch with a gasless abdomen suggesting the absence of a distal TEF.

In our series, a systematic screening of EA was carried out in 5 patients (24%). The total number of patients who were diagnosed within the first 24 hours was 13 (62%).

If esophageal continuity has not been verified at birth, infants with pure EA become symptomatic within the first few hours of life. Excess salivation and fine frothy bubbles in the mouth and sometimes the nose result from an inability to swallow. Any attempts at feeding result in choking, coughing, cyanotic episodes, and food regurgitation.

The presence of a fistula increases the risk of aspiration of gastric secretions into the trachea and lungs. Pneumonitis and atelectasis develop quickly in these neonates, and rattles heard during respirations are common. Fistulas also allow air to enter into the stomach and intestines, which can lead to abdominal distention. Gastric perforations occur, especially in the presence of imperforate anus. In the presence of atresia alone, the abdomen appears scaphoid.

The length of the esophageal gap is usually not known preoperatively. Absence of air in the stomach has been linked with a long gap, but has also been described in association with a distal fistula occluded with mucus [65]. Even in LGEA (atresia without distal fistula) as recently defined by the International Network of Esophageal Atresia (INoEA), the gap length can vary [5].

In newborns with isolated EA, the first procedure is generally a gastrostomy, which allows for enteral feeding and also allows for assessment of the length and location of the lower pouch. It can be identified radiologically, either with metal bougies, a small gastroscope, or with a contrast injected through the gastrostomy [66]. This can be done at the time of the initial gastrostomy or more routinely 7–10 days later. If bougies or a telescope are introduced into the distal esophagus, the amount of pressure on these instruments will affect the measurement of the gap between the two esophageal segments and may under- or overestimate the gap length. Bronchoscopy can also be performed prior to exploration, not only to assess the site of the distal fistula, but to also look for an upper pouch fistula. As previously mentioned, a proximal fistula has been found with a much higher incidence in cases of pure EA than previously reported, in one series up to 50% [67].

V. Associated malformations :

EA is frequently associated with other congenital malformations. The incidence of associated anomalies varies between 36% and 58% [68], [69].

In the study by Ghabili [70], whose aim is to compare the prevalence of VACTERL and non-VACTERL-type anomalies between patients with LGEA and those with non-LGEA, it was revealed that the prevalence of VACTERL spectrum anomalies was comparable between the two groups of patients. This finding is consistent with that of the study by Lopes [71]. However, non-VACTERL type anomalies were more common in patients with LGEA.

The rate of associated anomalies in our series is 38%. The highest rate was recorded in the series of Jensen et al. [1]. (Table XVII)

Table XVII : Comparison of associated malformations rate with other series

Series	Number of patients	Associated anomalies rate
Our series	21	38%
Series of Jensen el al. [1]	37	81,1%
Series of Stadil et al. [40]	71	61,4%
Series of Garabedian et al. [43]	88	56 ,7%

1. Cardiac malformations :

The most common associated malformations occur in the cardiovascular system with an incidence between 13% and 34% [39].These abnormalities seem to occur more frequently in patients with LGEA compared to those with non-LGEA [70], [72], [73].

Associated cardiovascular anomalies have a significant impact on the overall survival of infants with esophageal atresia, reducing the survival rate to 67% compared to 95% without cardiac anomaly [74].

The most common cardiac anomaly is the ventricular septal defect (19%), which is associated with an up to 16% mortality rate. Other common anomalies include atrial septal defect, tetralogy of Fallot, patent ductus arteriosus, aorta coarctation, transposition of the great arteries and dextrocardia. Some of these cardiac defects lead to a clinically evident heart insufficiency few days after birth and can be responsible for death before undergoing surgery. That is why it's crucial to screen these anomalies prior to surgery, on physical examination, chest X-ray and echocardiography.

Congenital vascular anomalies also have been associated with EA. The following anomalies have been described: right aortic arch (RAA), aberrant left subclavian artery (ALSA), aberrant right subclavian artery (ARSA) and double aortic arch [73].

Often asymptomatic, these abnormalities may be the cause of respiratory symptoms (dyspnea, cough, cyanosis) and/or may exacerbate gastrointestinal symptoms (dysphagia) when a ring completely or incompletely encircles the trachea and/or the esophagus, resulting in extrinsic compression.

Vascular defects are rarely diagnosed preoperatively on ultrasound or esophagram [73] and some authors recommand, when respiratory or digestive symptoms are present, the use of CT or MRI to screen for these anomalies [75].

2. Musculoskeletal malformations :

Reported rates of musculoskeletal anomalies in infants with EA range from 1,6% to 55% [76], [77]. The incidence of skeletal anomalies seems to rise when other atresias are also present. A 1976 paper by Gruchalski reported a 13.2 % incidence of skeletal involvement with esophageal atresia, this rose to 28,8% and 40% when there are anorectal atresia and anorectal + duodenal atresia respectively [78].

2.1. Limb anomalies :

The incidence of limb anomalies for the most part is not clearly delineated from that of general skeletal involvement in most published studies. In those that separated limb from spinal involvement, the incidence of limb anomalies in EA ranged from 8.9 to 42.7 % [77], [79], [80].

The most common form of limb anomaly is preaxial (radial) defect or deficiency (Figure 30), with an incidence of up to 35 % in VACTERL patients [79]. In a study utilizing pooled data from 11 birth defect registries, Rosano et al.[81] found that the odds of having a preaxial defect is 4.3 times higher in those with EA than those without.

Fernbach and Glass [82] specifically looked at limb anomalies other than preaxial abnormality in 24 VATER patients. They described an expanded spectrum of limb anomalies, including Sprengel deformity, humerus hypoplasia, radioulnar synostosis, midline hand anomaly, clinodactyly, and syndactyly.

Lower extremity involvement includes tibial field defect, hip dysplasia, congenital talipes equinovarus (Figure 31) and otherfoot deformities [79], [80], [82].

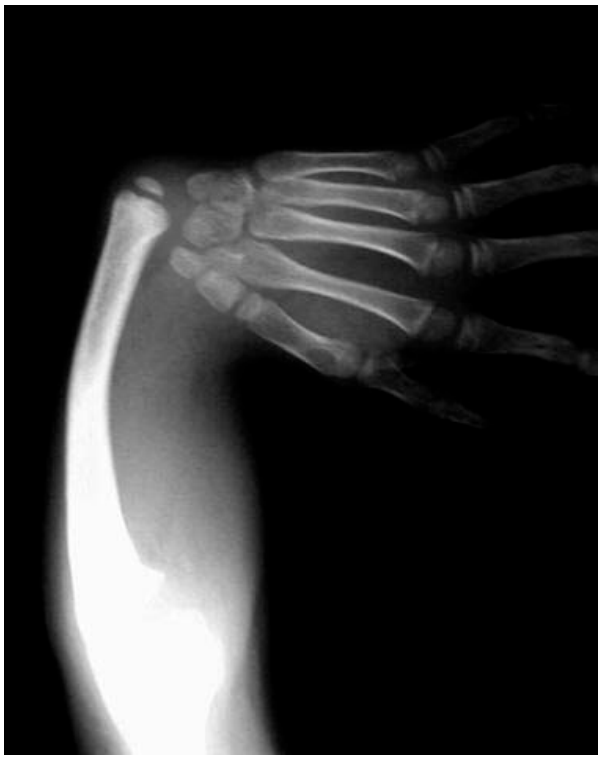


Figure 27 [4] : Radial clubhand (radial deficiency)

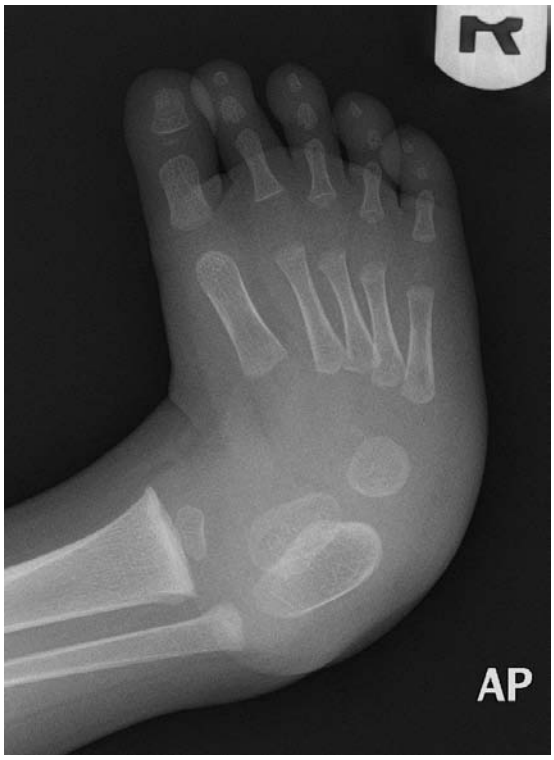


Figure 28 [39] : Congenital talipes equinovarus

2.2. Spine anomalies :

Reported incidence of spinal anomalies in EA ranges from 6.9% to 75% [77], [80].These include vertebral hyper- or hyposegmentation, abnormal number of ribs (Figure 29), hemivertebra, wedge or butterfly vertebra (Figure 30), unsegmented bar and congenital rib fusion.

The occurrence of tethered spinal cord (TSC) appears to be higher when esophageal atresia is associated with an imperforate anus or any urogenital anomaly [83].

An earlier claim that the presence of 13 pairs of ribs is a good indicator of LGEA has not been substantiated [84].

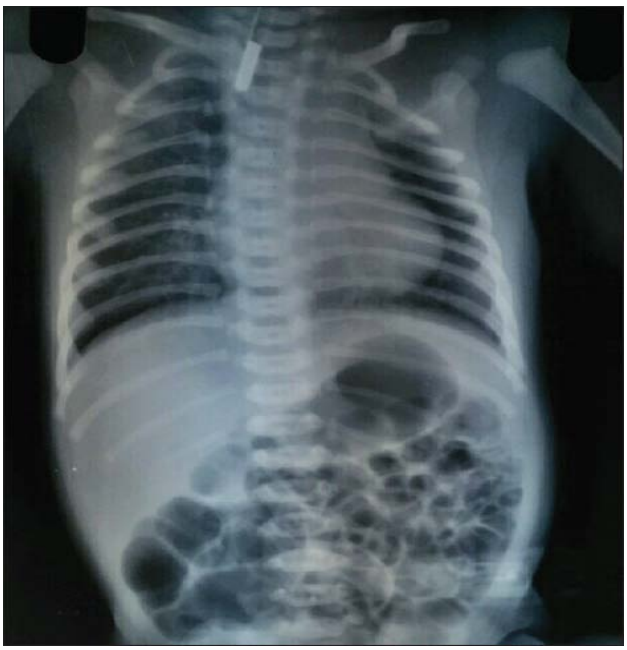


Figure 29 [85] :Associated Supernumerary ribs with EA

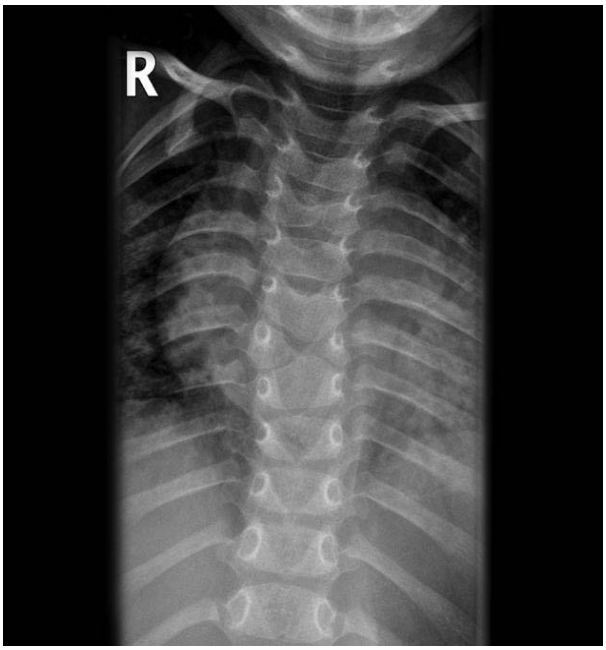


Figure 30 [86] : Butterfly vertebra

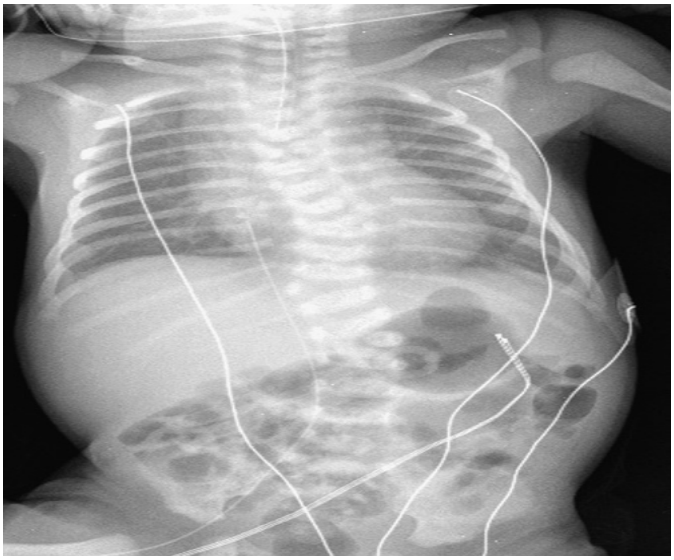


Figure 31 [87] :Associated scoliosis with EA

3. Genitourinary malformations :

The incidence of genitourinary malformations varies between 15% and 25% [14].

The majority of urinary tract abnormalities are incidental findings of no clinical significance such as ureteric duplication, unilateral agenesis and horseshoe kidney. Bilateral renal agenesis (Potter's syndrome) should be excluded on ultrasound scan in an infant who fails to pass urine. Vesico-urerteric reflux is the most common urinary anomaly which may or may not require active treatment.

Other urinary tract defects include polycystic kidneys, urethral atresia, urachal abnormalities, bladder extrophy [87].

The most common genital anomalies are hypospadias, cryptorchidism, ambiguous genitalia [87].

4. Gastrointestinal malformations :

Reported rates of digestive system defects in patients with EA range from 9% to 27% [14], [88]. Anorectal malformations account for the majority of the gastrointestinal defects with equal distribution between high and low anomalies.

Duodenal atresia has been reported in about 3% to 6% of EA patients [89]. It is a major etiology of congenital intestinal obstruction. The diagnosis is easily confirmed by the presence of the typical 'double bubble' sign on abdominal X-ray (Figure 32).

Another prevalent, but less well-known, associated malformation is Infantile Hypertrophic Pyloric Stenosis (IHPS). It has been described a 30 times higher prevalence of IHPS in patients with EA compared to the normal population [90].

The diagnosis should be considered when patients show recurrent or persisting vomiting and feeding intolerance after surgery. The increased prevalence of IHPS in EA patients suggests a relationship. However, no study has been carried out toward the cause of this increased prevalence. It is unclear if IHPS is the consequence of the surgical repair or the result of a shared genetic etiology.

The overall incidence of intestinal malrotation in patients with EA ranges from 3% to 5% [91]. This anomalie is more common in patients with EA type A [92]. Most malrotations remain asymptomatic but can evolve into a volvulus which is a lethal complication.

Other gastrointestinal defects include annular pancreas, diaphragmatic hernia, Hirschsprung's disease, Meckel's diverticulum, colonic and intestinal atresia.

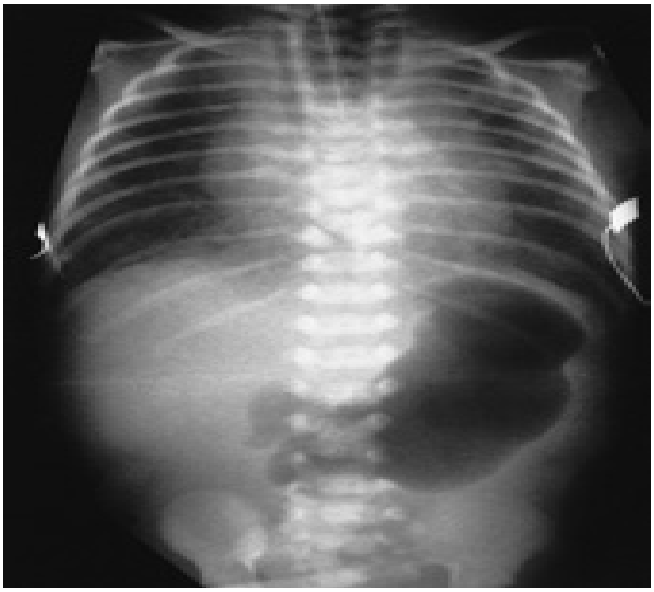


Figure 32 [14] : Double bubble sign confirms duodenal atresia in patient with EA.

5. Laryngotracheal malformations :

The prevalence of upper airway abnormalities in patients with EA was for a long time undervalued. Recent studies conducted for this purpose have shown that the prevalence of the association of these anomalies with EA reaches 40% [93], [94].

The most common defects are: tracheomalacia, vocal cord paralysis, subglottic stenosis, laryngeal cleft, respectively. Conforti et al. [93] have shown that mortality in EA patients significantly correlates with the presence of laryngotracheal abnormalities and recommend a systematic airway endoscopic preoperative evaluation to minimize laryngotracheal anomalies-related morbidity and mortality.

6. Chromosomal abnormalities :

The incidence of chromosomal anomalies averages 6-10% in patients with EA [95]. The most frequent anomalies are trisomy 21, trisomy 18, trisomy13, mosaic trisomy 8, triploidy, partial trisomies (dup 1q32-qter, dup 2q,dup 5q, dup 7p and dup 12q) [95]. A carytotype is then justified in case of malformative association or in presence of facial deformities[96].

7. Non VACTERL-type malformations :

These malformations include :

- Lower respiratory tract anomalies : Usui et al. [97] reported a 47% incidence of tracheobronchial abnormalities such as ectopic or absent right upper lobe bronchus, congenital bronchial stenosis, and a decreased ratio of circumferential cartilaginous trachea tomembranous trachea.
- Nervous system anomalies : Hydrocephaly, microcephaly, hypoplasia corpus callosum, spina bifida [75], [96].
- Craniofacial anomalies : cleft palate, choanal atresia, zygomatic arch cleft, ear deformities.
- Dermatological anomalies : skin anomalies occur in 21% of patients born with with LGEA [75].

8. Malformative associations :

In 1973, Quan and Smith [98] suggested a broad spectrum of associated malformations that are associated with EA malformations. They arranged this association into the acronym VATER: Vertebral defects, Anal atresia, Tracheoesophageal fistula, Esophageal atresia, and Renal defects. Opitz defined the term “association” as the idiopathic occurrence of multiple congenital anomalies during blastogenesis. Affected patients have no family history of malformations, no

recognizable teratogen is involved, and no chromosomal abnormalityis observed. As the phenotype expanded, the acronym was changed to the VACTERL association, which includes Vertebral, Anorectal, Cardiac, Tracheoesophageal, Renal, and Limbabnormalities [99]. The incidence of the VACTERL association in the EA population is approximately 20%. This association seems to be less common in patients with LGEA [11]. The diagnostic is confirmed if 3 or more of the above anomalies are present.

Infants with EA malformations in association with the VACTERL association have a high mortality rate. Driver et al. [100] reported that the relative risk for mortality in patients with VACTERL associations was 2.54.

The other reported malformative associations reported in EA patients are :

- CHARGE association : The components of the CHARGE association comprise Coloboma (85 %), Heart defects, choanal Atresia, Retarded growth and development, Genital hypoplasia (almost all boys) and Ear deformities. Around 2 % of infants with EA have this association, while of infants with the CHARGE association, 16 %have EA [101], [102].
- Potter's syndrome : Potter's syndrome comprises renal agenesis, pulmonary hypoplasia and typical facial dysmorphia. Three quarters of infants are male [88].
- Schisis association : it comprises omphalocele, cleft lip/palate and genital hypoplasia [103].
- Pierre Robin syndrome : this syndrome comprises mandibular hypoplasia, glossoptosis and occasional mental retardation [88].
- Feingold syndrome : Feingold syndrome is characterised by autosomal dominant inheritance of microcephaly and limb malformations, notably hypoplastic thumbs and clinodactyly of second and fifth fingers [88].
- Fanconi syndrome : Hypoplastic thumb (80 %) and/or radial hypoplasia or complete absence of the radial bone, pigmented skin lesions, small penis, mental retardation in some, pancytopaenia and lymphoreticular malignancy [88].

Cardiovascular defects followed by musculoskeletal malformations are the most frequent in our series, which is consistent with the majority of series in Table XVIII. Genitourinary malformations come in 3rd place in our series, which is similar to the results of the other series with the exception of the series of Bardain et al. The gastrointestinal defects were seen in 10% of our patients, the highest rate was recorded in series of Bairdain et al. Karyotype was not performed in any of our patients, therefore the prevalence of chromosomal abnormalities could not be evaluated. In the other series, the prevalence of chromosomal defects ranges from 2,7% to 21%. VACTERL association was recorded in 5% of our cases, which is comparable to the series of Holland et al. The highest rate of VACTERL association was recorded in series of Bairdain et al. (Table XVIII)

Table XVIII : Distribution of associated malformations in our series compared with other series

	Our series	Series of Bairdain et al. [11]	Series of Holland et al. [20]	Series of Gallo et al. [18]	Series of Jensen [1]
Cardiovascular defects	19%	44%	6,19%	25%	59,4%
Musculoskeletal defects	19%	25%	4,13%	29%	37 ,8%
Gastrointestinal defects	10%	30%	2,7%	12,5%	27%
Genitourinary defects	14%	25%	5,16%	21%	29,7%
Chromosomal defects	-	13%	2,7%	8%	21%
VACTERL association	5%	25%	6%	-	-

VI. Therapeutic management :

1. Preoperative management :

1.1. Conditioning :

Once the diagnosis of EA has been established, the baby, if not delivered in a maternal/neonatal center, should be transferred to a regional pediatric surgical center with intensive care support facilities. Adequate and immediate management with the following measures must be undertaken :

- Patient is nursed with the head elevated (head-up at 30° to 45°) to reduce the the risk of pulmonary aspiration [104]. An upright sitting position has traditionally been advocated, but some authors argue that the head-up, prone position is most effective at minimizing reflux [87].
- A French Replogle tube is placed in the upper esophageal pouch (0,5 cm above the end of the pouch) and connected to continuous low pressure suction of -11 to -25mmHg. For infants greater than 1500g a 10Fg tube is used. The tube should be inserted nasally when possible ; if not, then orally. For infants below 1500g, a 8Fg tube is recommended.
- Routine endotracheal intubation should be avoided unless there is a respiratory distress, severe pneumonia or severe associated malformations that require respiratory therapy. Mechanical ventilation increases the risk of gastric distension and rupture of the stomach due to the passage of a large amount of air through the TEF. This can be prevented by positioning the tip of the tracheal tube beyond the fistula and using a low-pressure ventilation. When this is not possible (in case of fistula next to the carina), the fistula can be occluded with a Fogarty catheter. Although, most of these cases require the ligation of the fistula in emergency [104].

- Intravenous access should be established to infuse patients with 10% dextrose and hypotonic saline in order to maintain fluid, elecrolytes and glucose balance. Vitamin K analogue should also be adminitrated. Oral feeding should be prohibited [87].
- Preoperative antiobioprophylaxis is still debated. Some authors restrict its administration to patients in whom oral feeding has already been initiated and to patients with clinical and/or paraclinical signs of infection [64], [105]. While other authors recommend that broad-spectrum antibiotic therapy should be administered routinely [14], [87].
- Premature newborns are subjected to cooling during transport from the delivery room to intensive care and during diagnostic tests; for this reason they should be placed in an incubator or on a preheated surgical bed, and protected with devices to limit the heat dissipation [104].
- It is also essential to identify the pain (through facial expressions and postures and by changes of vital signs) preventing it with analgesics and reassuring the child; crying causes an increased passage of air through the tracheo-esophageal fistula with increased gastric distension, elevation of the diaphragm and impaired respiratory dynamics [104].
- Cardiorespiratory monitoring [39].

1.2. Preoperative assessment :

The two primary goals of preoperative assessment in patients with a clinical diagnosis of EA are: confirmation of the diagnosis, identification of associated anomalies with immediate management implications for the planned EA surgery.

a. Routine imaging :

Associated malformations are screened by :

- **Complete physical examination** : a careful clinical examination should be conducted in order to rule out associated abnormalities such as: facial dysmorphies, anorectal

anomalies, limb and ribs defects, pathologic heart murmur and absent femoral pulses, ambiguous genitalia. These associated anomalies are mainly responsible for the medium- and long-term prognosis in these patients.

- **Thoracoabdominal X-ray** : confirms the diagnosis and shows sign of a distal TEF (abdominal gas) if it is present. It evaluates the existence of cardiac silhouette abnormalities in favor of cardiac defects and an absent left aortic countour suggesting a right aortic arch [106]. The presence of rib and vertebral malformations, duodenal atresia (double bubble sign) and diaphragmatic hernia can also be evaluated.
- **Echocardiography** : evaluates cardiac and vascular anomalies. According to ERNICA (European Reference Network for Rare Inherited Congenital Anomalies) consensus conference on the management of patients with LGEA 2019 [107], echocardiography should be routinely performed as a preoperative diagnostic procedure, especially to exclude a right aortic arch and a right descending aorta which influences the choice of the surgical approach. More recent techniques to determine the side of the arch include computed tomography angiography (CTA) and magnetic resonance angiography (MRA), CTA permits exceptional visualization of the vascular anatomy and has the added benefit of three-dimensional reconstruction [108]. MRA offers similar cross-sectional imaging advantages to CTA, but without the risk of ionizing radiation [109]. Due to technical demands, transport requirements and anesthesia, it is unlikely to routinely employ these methods in the clinical setting. The most satisfactory method continues to be echocardiography, which can be performed at the bed side without radiation exposure and can also evaluate significant cardiac defects.
- **Abdominal ultrasound** : allows to assess gastrointestinal and urinary tract defects. Its realization preoperatively is still debated [107].

b. Tracheobronchoscopy :

It defines the anatomy of the respiratory tree, confirms the presence of a proximal and/or distal TEF; the site of entry and location. The value of routine preoperative rigid tracheobronchoscopy is much debated because the incidence of a simultaneous proximal and distal fistula is <5% [66]. However, two studies reported that the routine use of tracheobronchoscopy led to detection of a much higher relative incidence of proximal fistula, up to 5.69% [67], [110]. Tracheobronchoscopy allows also a clear vision of the entrance of the distal fistula (Figure 33), its distance to the carina provides a clue as to the length of the gap between the esophageal segments: the closer the fistula is to the carina, the longer the distance between the esophageal segments [111]. It may also reveal abnormalities such as tracheomalacia, a laryngotracheoesophagealcleft (Figure34), tracheal stenosis, or a tracheal bronchus to the right upper lobe. With the availability of small-diameter flexible fiberscopes, it can now be performed after intubation through the endotracheal tube [112]. However, forceful ventilation must be avoided, not only to avoid lung damage [113], but also to prevent gastric distention and gastric perforation with insufflations through the distal fistula.

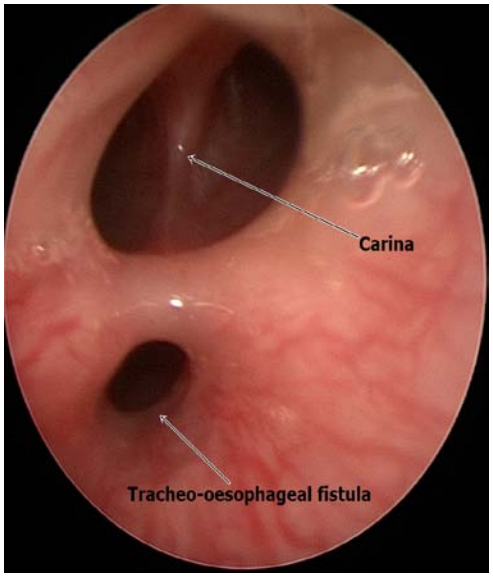


Figure 33[114] : Bronchoscopy of a distal TEF.

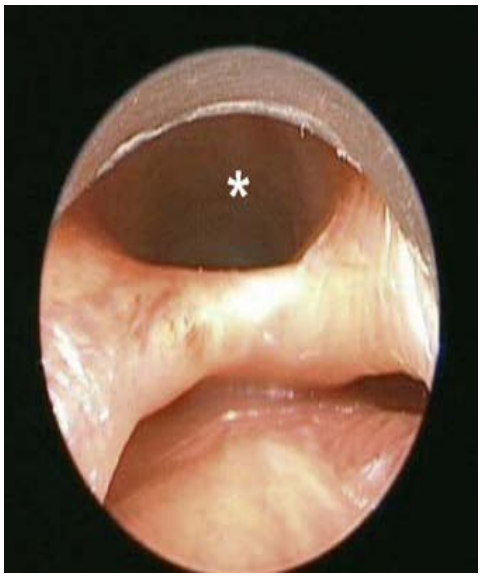


Figure 34 [39] : Bronchoscopy of a laryngotracheoesophageal clif with the trachea anteriorly (asterix) and the esophagus posteriorly.

c. Other investigations :

- **Computed tomography (CT) :** The routine use of preoperative CT scans in newborns with EA is controversial as the limited information acquired that may help to change the surgical plan can be easily obtained by tracheobronchoscopy or intraoperatively. Su et al[115] found no differences in the distance between the two esophageal pouches as measured by CT scan and at surgery, and the same results were achieved by Wen et al[116] by utilizing multidetector-row computed tomography (MDCT) in reconstruction of 3D volume rendering. A recent review of the 8 available studies on the topic suggests that the safety of CT scan techniques is questionable due to limited facilities, problems regarding neonatal transportation to the radiology department and the need for sedation. Moreover, although a modern CT gives low grade exposure, this examination is still associated with radiation hazards [117].
- **Magnetic resonance imaging (MRI) :** The experience with MRI in newborns affected by EA is extremely limited [67]. Cantinotti et al. [118] consider this method to be an important diagnostic tool in identifying anomalies of the aortic arch and associated cardiac anomalies. Nevertheless, the advantages of the visualization of tracheobronchial and esophageal system have not been sufficiently demonstrated, and the need for general anesthesia makes magnetic resonance imaging a procedure only for selected cases.
- **Spine X-ray :** should be performed if a spinal abnormality has been suspected on physical examination and on the initial thoracoabdominal X-ray.
- **Karyotype :** is indicated if there are several associated malformations with facial dysmorphia suggestive of a particular chromosomal anomaly [63].
- **Cranial ultrasound :** indicated especially in premature newborns to diagnose intraventricular hemorrhage and periventricular leukomalacia. It's also performed in case of macrosomia to check for congenital hydrocephalus.
- **Blood test :** is routinely carried out. It includes : CBC, hemostasis test, CRP, BUN, creatinine, serum electrolyte test, arterial gasometry, ABO rhesus typing, transaminases and bacteriological samples.

2. Surgical management :

2.1. Aims :

- In first stage, perform a gastrostomy to allow enteral feeding and the 2 segments of the esophagus to grow. A closure of the TEF is also carried out in patients with EA type C.
- In second stage, re-establish the continuity of the esophagus either by preserving the native esophagus (delayed primary anastomosis) or by esophageal replacement (esophagoplasty).

2.2. Surgical approaches and procedures :

The operative approach to an infant with LGEA depends greatly on the specific type of anomaly present and the occurrence of associated anomalies.

a. Initial gastrostomy :

Placement of the gastrostomy tube is the first operative phase in the treatment of LGEA and it allows the early institution of enteral feedings, which leads to subsequent enlargement of the diminutive stomach and stretching of the distal esophageal pouch. Enlargement permits the stomach to be used as an esophageal substitute if necessary [87].

Several approaches have been described for establishing a gastrostomy ; the most widely deployed is the Stamm technique (gastrostomy with laparotomy) : the stomach is approached through a short, transverse supraumbilical incision (Figure 35). Fascial layers are incised transversely and the muscle is retracted or transected. The catheter exit site should be approximately at the junction of the lower two thirds and the upper one third of a line drawn from the umbilicus to the mid portion of the left rib cage, over the mid rectus muscle. It can be useful to mark the site prior to making the main incision. A vertical incision is used in children with a high-lying stomach or a very narrow costal angle [119].

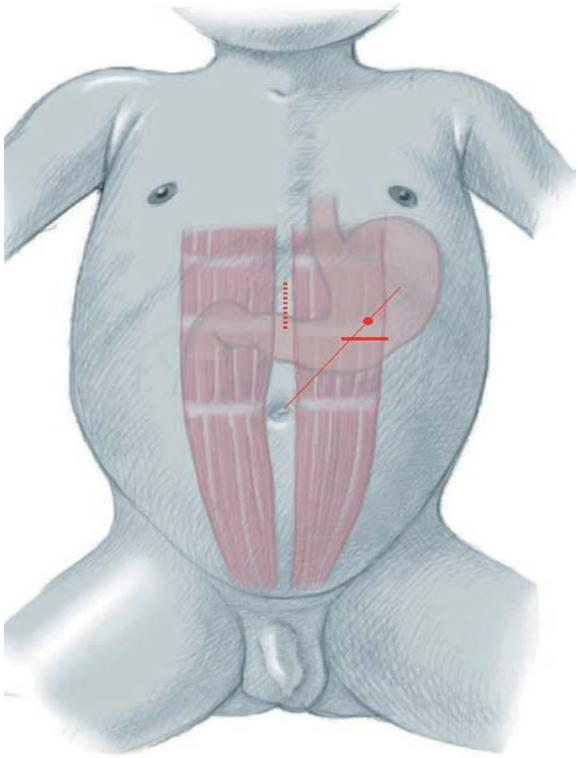


Figure 35 [119] :Transverse supra--umbilical incision

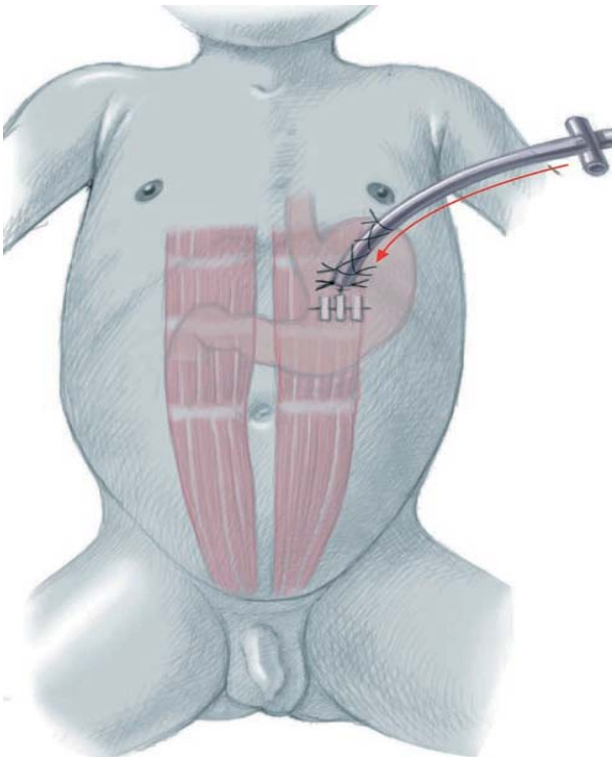


Figure 36 [119] : Gastrostomy tube fixation

Less invasive techniques for gastrostomy establishement have been reported to be feasible and safe in the pediatric population, including the percutaneous image-guided gastrostomy. The advantages of this technique are avoidance of a laparotomy and, in some cases, elimination of the need for general anaesthesia [120]. (Figure 37)

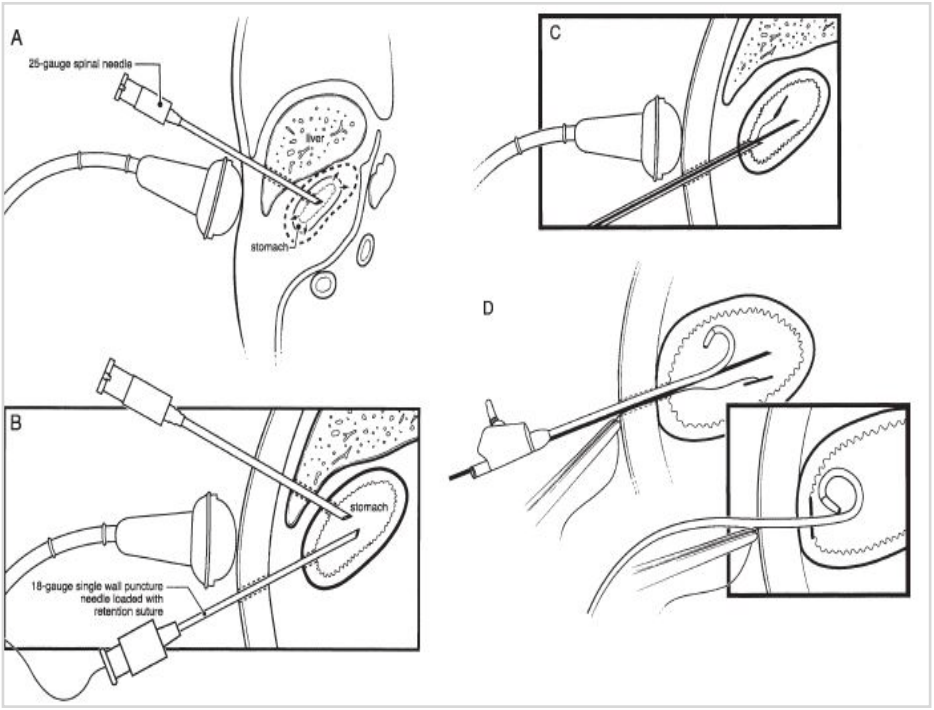


Figure 37[120] : Diagram shows the technique for image-guided percutaneous gastrostomy insertion in a child with pure esophageal atresia.

(A) The stomach is identified sonographically and accessed with a 25-gauge spinal needle going through the left lobe of the liver. Air is injected to distend the stomach.

(B) An 18-guage single wall puncture needle loaded with a retention suture is then passed through the abdominal wall into the distended stomach and its location confirmed with water soluble contrast.

(C) A 0.035-inch wire is inserted, which deploys the retention suture into the stomach.

(D) The needle is withdrawn, leaving the wire in place. The tract is dilated, and the catheter is inserted.

b. Tracheoesophageal fistula ligation :

Patients with LGEA type C undergo, in addition to the initial gastrostomy, a repair of the TOF.The standard approach is a right laterodorsal thoracotomy, with the patient placed in left lateral decubitus (right side up) with the right arm extended and a roll placed under the left chest [4]. (Figure 38)

The azygos vein represents a major landmark in any TOF repair (Figure 39). Dissection of the vein may not be necessary, but it certainly facilitates an easier identification of the TEF. Next, the vagal nerve fibers should be identified as they converge to the distal esophagus. The distal esophagus is encircled with a vessel loop to permit an atraumatic mobilization [88]. (Figure 40)

Usually, the distal pouch is more hypoplastic than the upper pouch, and extensive dissection is not recommended in order to preserve the blood supply and vagal innervation. Instead, the TEF is mobilized straight forward to its junction with the trachea. Especially, in preterm babies, this preparation should be performed with great care to avoid damaging the membranous portion of the trachea. The TEF should be divided close to the tracheal wall and leave 1mm or 2mm of esophagus on the tracheal end of the fistula to minimize the risk for postoperative tracheal stricture, while not leaving an excessive amount that would act as tracheal diverticulum [87]. This cuff is closed with interrupted 4-0 or 5-0 sutures [88]. (Figure 41)

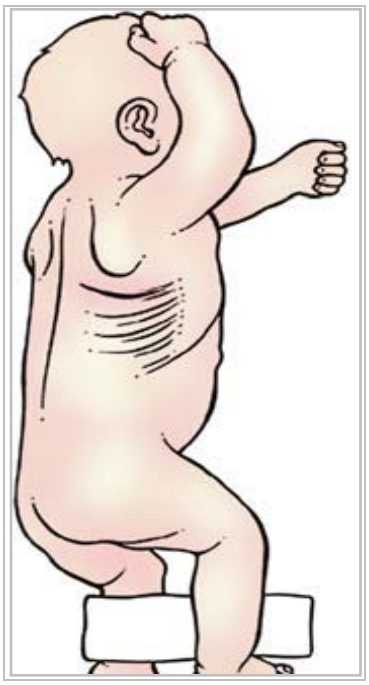


Figure 38 [39] : Patient positioning for right posterolateral thoracotomy

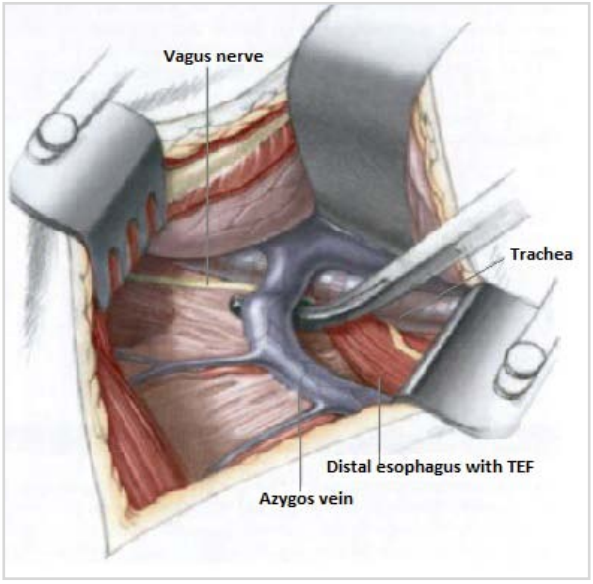


Figure 39 [88] : The azygos vein represents a major landmark of TEF repair.

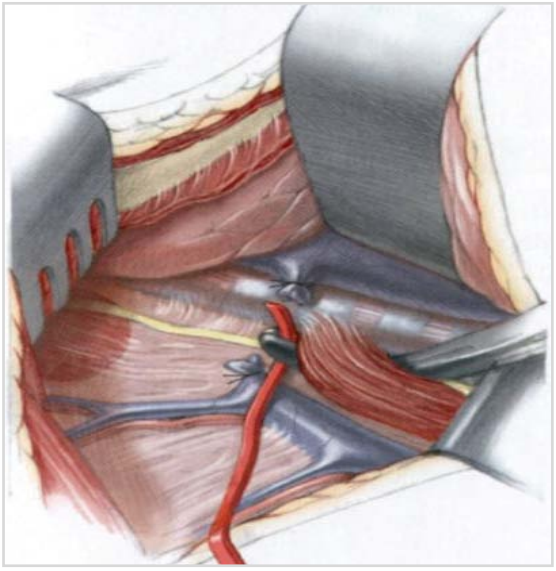


Figure 40 [88] : The TEF has been encircled with a vessel loop to allow atraumatic preparation. Vagal nerve has been preserved

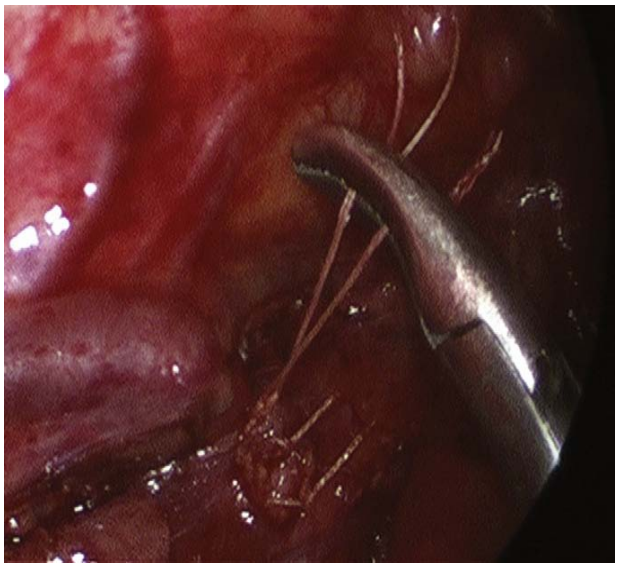


Figure 41 [87] : Interrupted suture closure of TEF

c. Esophageal lengthening techniques :

Several techniques have been described, additionally to the swallowing reflex and gastric reflux, as a stimulus for the growth of the 2 segments of the esophagus in order to shorten the time till the anastomosis is possible.

c.1. Longitidunal bougienage :

The bougienage of the upper pouch has first been reported by Howard and Myers in 1965. Many further publications followed supporting this technique (Mahour et al. [121]). In 1966, Lafer and Boley [122] reduced the waiting period to 6 weeks by bougienage of the upper and lower esophagus. Technical modifications have been published by Hendren and Hale [123] using electromagnetic devices for the longitudinal bougienage. (Figure 42)

Recent experience shows that daily bougienage of the upper and lower pouch for a few minutes within the incubator and under light sedation allows to achieve an overlap of the segments on the X-ray within 3-4 weeks until the anastomosis can be performed (Figure 43). Although not widely accepted as a useful method, the experience shows that the time span till an anastomosis is performed can be significantly reduced and secondary resection are rarely needed.

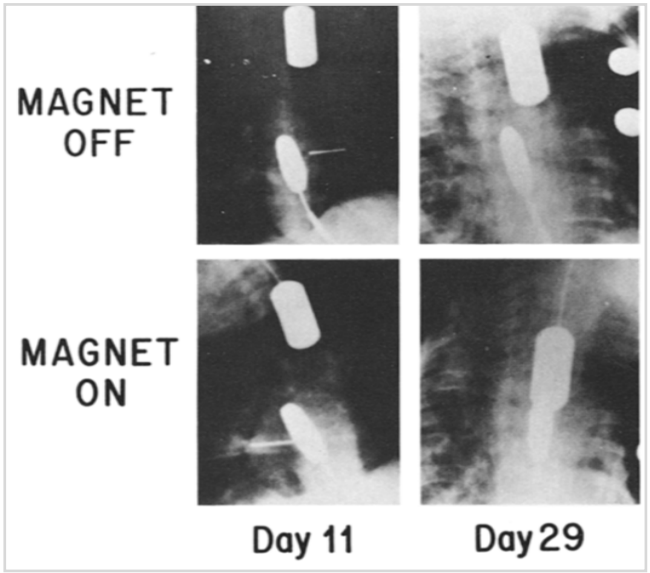


Figure 42 [123] : Roentgenograms on two occasions are shown with the magnet off and on. The gap between bullets is less with the electromagnetic field activated. A gap of 3-4 cm closed from day 11 to day 29.

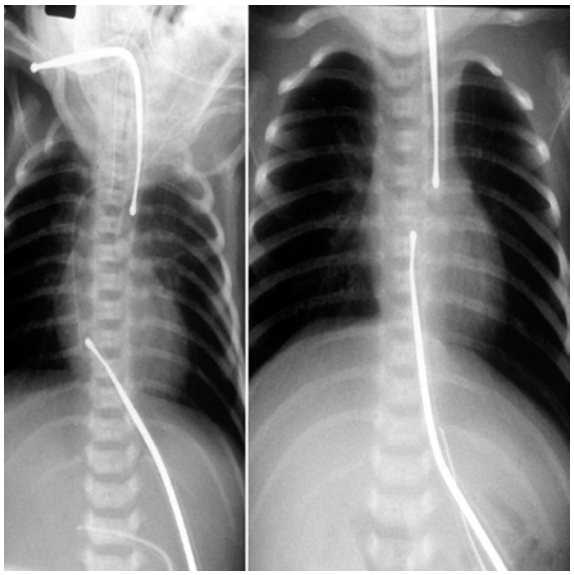


Figure 43 [14] : Longitidunal bougienage of the upper and lower pouch (metal probes within plastic tubes). A near overlapping of the esophageal segments was achieved within 3 weeks.

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c.2. Forced traction techniques :

Extensive tension at the anastomosis can lead to severe leakage or even disruption of the esophageal segment. To circumvent this problem, but still to be able to perform an anastomosis and preserving the child's own esophagus, forced traction methods have been developed.

Rehbein[124] was the first to publish a method of intraesophageal forced traction by means of silver olives in eachsegment which are pulled together along a nylon thread. A nearly identical procedure with Teflon balls instead of silver olives was proposed by Harrison [125]. Originally the thread was inserted into the esophageal segments by thoracotomy, later this procedure was performed endoscopically by Booss et al.[126]. The aim of the technique was to bring the two segments together and to create an autoanastomosis within 10 days. The resulting stenosis needed long-term bougienage and in 50% secondary resection. Recently, Stringel et al. [127] developed a similar technique which consists in creating an autoanastomosis by means of a thread was successfully applied in five patients.

In contrast to the internal traction method, an external traction technique was introduced by Foker et al. [128]. During thoracotomy, tissue-pledgetted traction sutures are placed extramucosally in the upper and lower segment and brought out to the skin below and above the incision (Figure44). Daily external traction of these sutures brings the segments together within 14 ± 2.9 days and the anastomosis can be performed by a second thoracotomy.

Recently, even a thoracoscopic elongation of the esophagus was successfully performed by Van der Zee et al. [129], thus avoiding the two routine thoracotomies. According to Foker, the technique successfully elongates esophageal segments which are separated even by ultra-long gaps up to 12 cm. But the complication rate is significant : in 28.5% of 42 patients additional rethoracotomies were needed due to pulled out traction sutures, replacement of traction sutures, or adhesions, and in two out of ten patients a secondary resection of the anastomosis was performed due to stenosis [128]. A survey [130] of 88 international surgeons showed that 39% are using the Foker technique, but 24% of those were not satisfied with the results. However,

Oliver et al. [131] have recently shown promesing results with low peri-procedural morbidity and favorable functional outcomes in 9 patients who underwent Foker procedures.

In 1994, Kimura and Soper [132] developed a technique of extrathoracic elongation of the upper pouch (Figure 45). First, a right-sided esophagostomy is created in patients with a LGEA. This stoma is advanced subcutaneously after 2–6 months several times until the proximal esophagus is long enough. In 12 patients, a final anastomosis was possible after 30 months (range 13–61 months); none of them needed a rethoracotomy or secondary replacement. Tamburri et al. [133] performed the procedure in 3 patients thoracoscopically.

Experiences of combining the Foker technique with Kimura's method resulted, however, in a high complication rate [134].

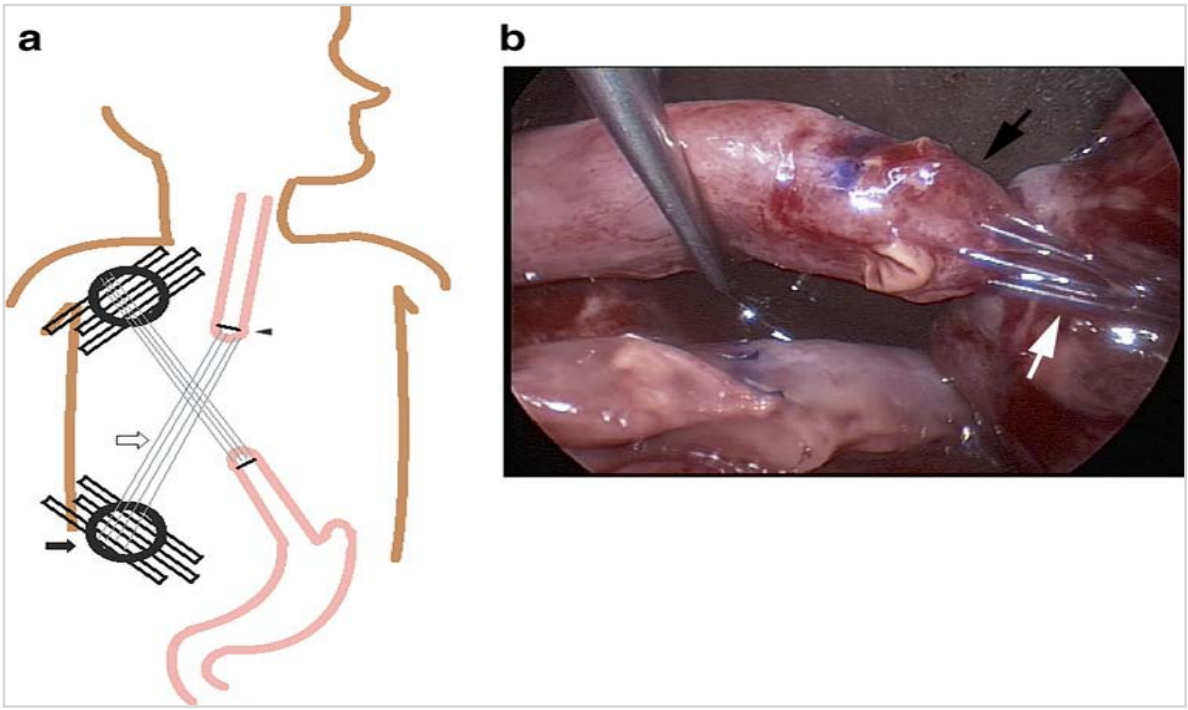

Figure 44 [135] : Foker procedure.

(a) Schematic demonstrates esophageal segments marked by radiopaque clips (black arrowhead) and attached to lenghtening apparatuses in the chest wall (black arrow) with traction sutures (white arrow).

(b) Intraoperative picture demonstrates an upper esophageal pouch (black arrow) with traction sutures in place (white arrow).

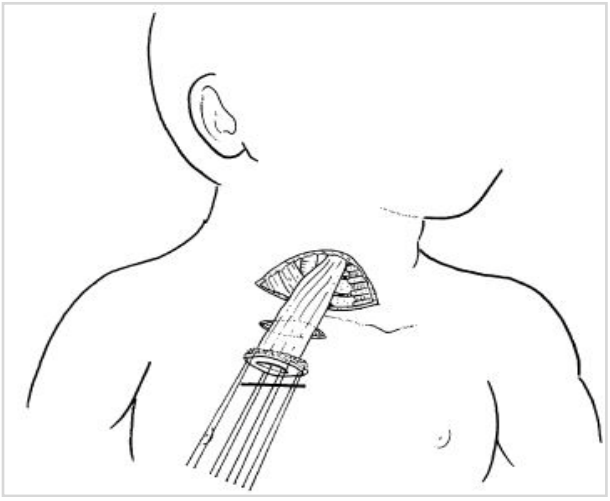

Figure 45 [136] : Kimura procedure

c.3. Myotomy :

Esophageal myotomy can be considered as an adjunctive maneuver to bridge a 1–2 cm intraoperative gap [137]. Both circular myotomy and spiral myotomy have been described to obtain length on the native esophagus to allow for a primary repair. These efforts have traditionally focused on lengthening the proximal pouch because it is felt to have a more robust blood supply leading to a safe anastomosis. Lai et al. [138] described a series of 5 patients who underwent circular myotomy of both the proximal and distal esophagus. The length of the gap in these patients measured 4.5 cm to 6.5 cm.Two patients had anastomotic leaks, one developed a stricture, and four patients developed reflux. All patients were found to have mucosal outpouching (diverticulum) postoperatively, but all retained normal esophageal function and were able to tolerate full oral feeds. The authors suggest that distal myotomy is safe and effective in the management of LGEA [138].

Beger and Beger [139] conducted a in vitro experience on 21 esophagus samples from 21 male lambs. The aim of the study was to review V-myotomy (VM) technique and the differences of the said technique with Livaditis circular myotomy (LM) and Kimura spiral myotomy (KM) techniques. VM was found to signifcantly increase total esophagus length and elongation per incision over LM and KM. In addition, VM was also shown to have a higher perforation pressure. (Figure 46 and 47).

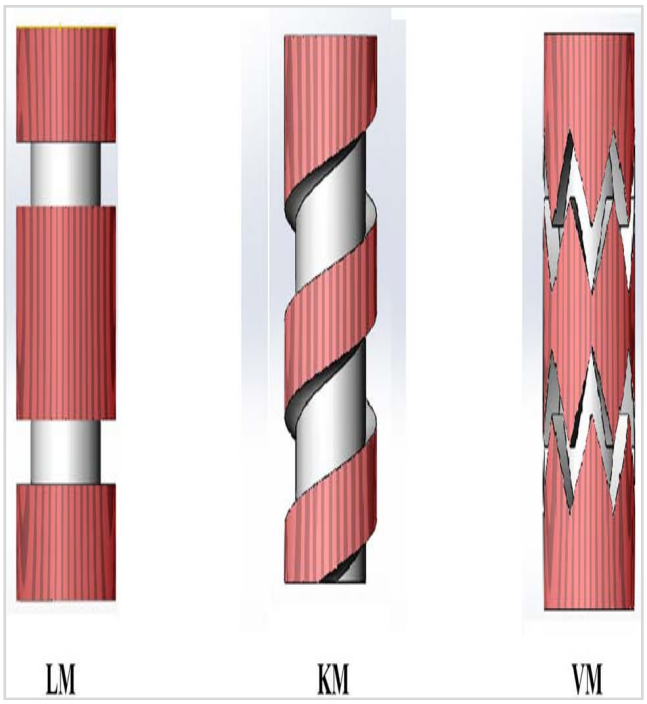

Figure 46 [139] : Techniques of LM, KM and VM.

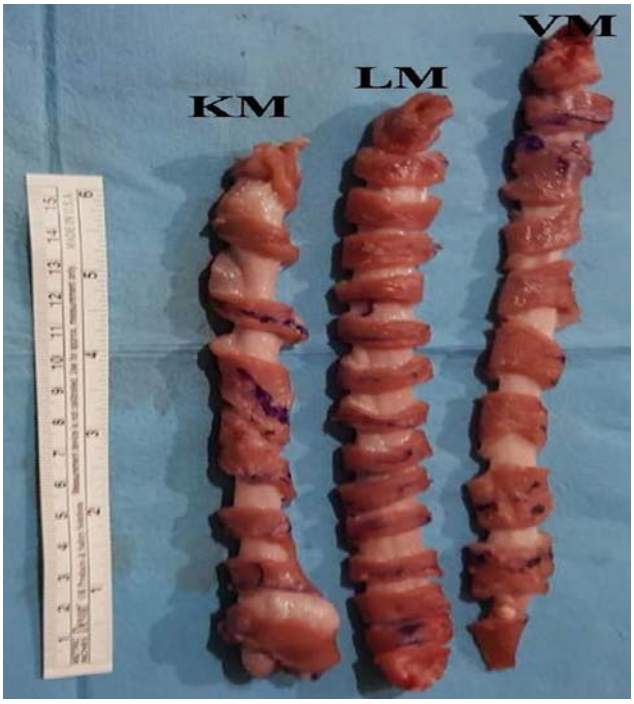

Figure 47 [139] : Techniques used in samples and their final lengths.

d. Definitive repair of LGEA :

d.1. Delayed primary anastomosis (DPA):

Efforts should be directed to preserving the native esophagus whenever it is possible because an ideal graft does not exist. Therefore DPA is considered the best early option for LGEA [137].

Puri et al. [140] have shown that the maximal spontaneous growth of the two esophageal segments occurs in most patients by 8–12 weeks of age, and this correlates with doubling of birth weight. By this age, the gap between the two esophageal pouches usually is less than 2 cm [141], [142]. Therefore, it is recommended to perform DPA when the patient is 3–4 months old. Successful primary anastomosis with delays of up to 12 months[143] and initial gaps of up to 7 cm [144] or 8 vertebral bodies [145] has been reported.

The surgical approach for DPA of the esophagus in LGEA is similar to that of repair of EA with TEF. A standard right posterolateral thoracotomy is performed via the fourth intercostal space using the extrapleural approach. Unless a right aortic arch is identified preoperatively, then a left-sided thoracotomy is prefered. A modified posterior thoracotomy, which preserves the latissimus dorsi and the thoracodorsal nerve, with satisfactory exposure, functional, and cosmetic results has been recently described as a reliable approach in the treatment of EA patients [146].

The advantage of the extrapleural approach is that a postoperative anastomotic leak does not contaminate the pleural cavity, allowing prolonged chest-tube drainage [88]. (Figure 48)

Friedmacher et al. [147] and Stadil et al. [148] have found that GER was more common in patients who underwent a DPA compared to other techniques. This can be due to the mobilization of the distal esophagus and the displacement of the gastroesophageal junction upward to the thoracic cavity. However, survival rates as well as long-term results are excellent when compared with other methods [141], [149].

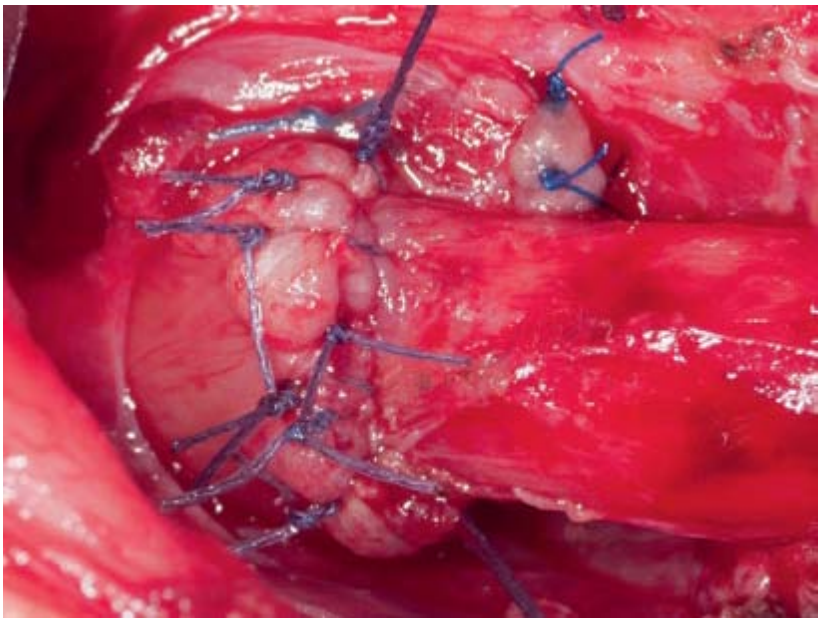

Figure 48 [88] : DPA with interrupted sutures and extraluminal knots

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d.2. Esophageal replacement :

Esophageal replacement is considered after an unsuccessful traction procedure or a failed primary repair [137]. It is generally accepted that the ideal esophageal replacement conduit for children should be long-lasting, be associated with minimal reflux, be technically feasible, not affect cardiac or pulmonary function, and allow oral consumption of nutrition. Four options have been described for esophageal substitution: gastric transposition (gastric pull-up), gastric tube, colonic interposition and jejunal interposition.

- **Gastric transposition:**

In children who require esophageal replacement, gastric transposition has been well described in the literature. Several studies reviewed the use of gastric transposition as an esophageal subtitute for LGEA [150]-[153]. Sharma and Gupta [150] reported the use of gastric transposition in the neonatal period in 6 patients. The mean length of stay was 14,6 days and all patients were able to tolerate oral feeding. Ng et al. [153] compared the use of laparoscopic approach to open approach for gastric tube in infants. No difference was found in anastomotic leaks rates, stricture rates and mortality rates between open and laparascopic techniques. A survey of European pediatric surgeons [154] and a review by Von Allmen et al. [155] showed that in cases where an esophageal replacement was needed, gastric transposition was the preferred procedure. (Figure 49)

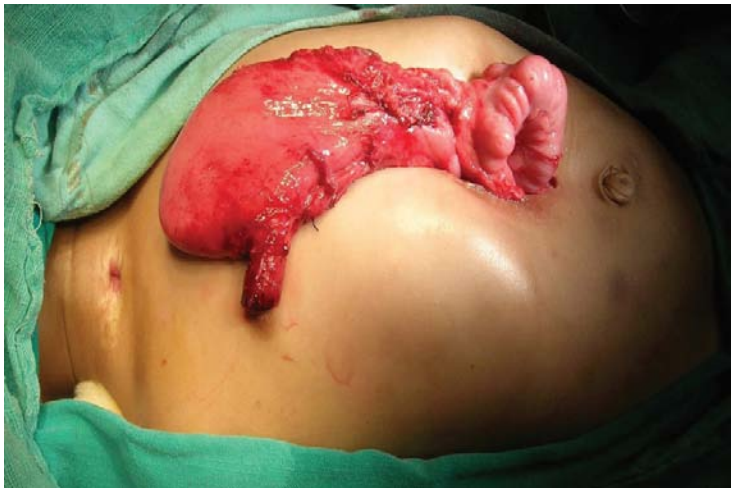


Figure 49 [156] : Intraoperative image of a gastric transposition.
Stomach fully mobilized and easily reaching the neck.

- **Gastric tube:**

The use of gastric tubes has evolved from the classic "reverse" gastric tube (Figure 50), which creates an antiperistaltic conduit to modifications allowing for an isoperistaltic tube (Figure 51), and the Scharli technique that uses the lesser curve of the stomach. Although complication rates are high and the technique is much less common than the gastric transposition, a recent series was published by Gupta in which a total of 16 children were evaluated of whom 10 had a history of EA. Overall, 14 of the 16 were eating and swallowing normally and 12 were between the 3rd and 97th percentile for weight. By report, almost all were asymptomatic and led a normal life [155].

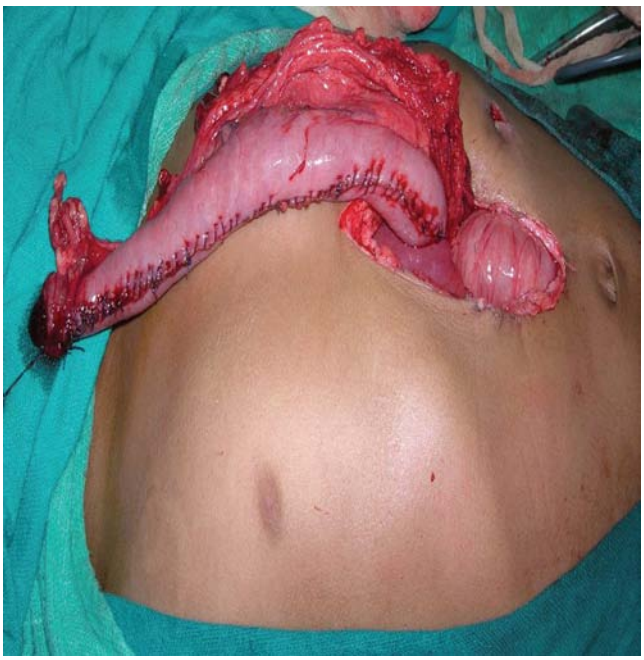


Figure 50 [156] : Intraoperative image of a reverse gastric tube.Well vascularized graft with adequate length.

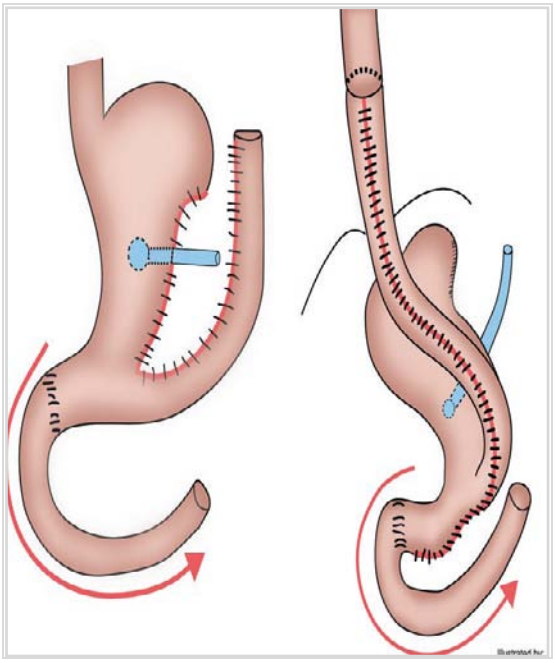


Figure 51 [88] : The iso-peristaltic gastric tube illustration

- **Colonic interposition :**

Use of the colon as a conduit to replace the esophagus was first described in 1948 and it remains one of the standard techniques used to bridge the gap in LGEA. The procedure employs either the right or left colon with a vascular supply frequently based on the middle colic artery

perfusing the marginal artery. The graft can be placed in either a peristaltic or antiperistaltic orientation and sufficient length can be obtained to reach the pharynx if necessary. The interposition can be placed in the posterior mediastinum or substernally. Classically, the distal end of the interposition is anastomosed to the antrum of the stomach, but segmental interpositions have been described that connect the proximal and distal segments of the esophagus to preserve the gastroesophageal junction and limit the length of the graft [155]. (Figure 52)

Tannuri et al. compared gastric transposition and colon interposition in a single center over 27 years. Colon interposition was carried out in 115 patients while gastric transposition was performed in 34. There were no differences in graft necrosis rates (one in each group) or mortality. The authors noted a higher rate of minor complications in colonic interposition with a lower rate of major complications. They concluded that both techniques were suitable for esophageal replacement; however, they recommended colonic interposition as the first choice.

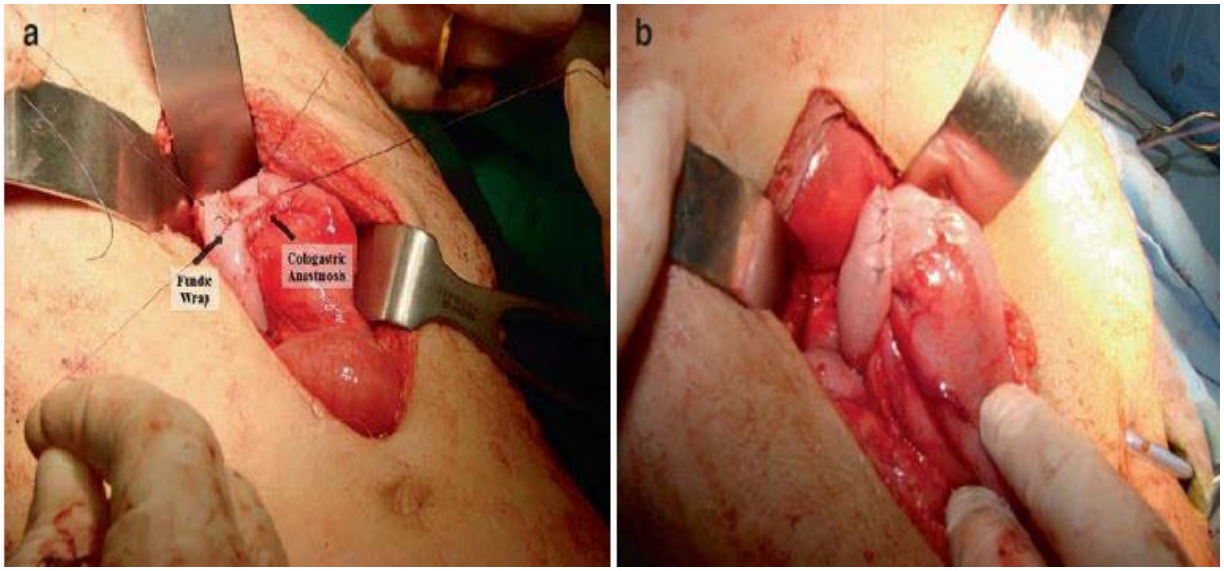


Figure 52[88] : Antireflux 270° around the cologastric anastomosis

- **Jejunal interposition :**

Use of a segment of jejunum, as an surgical option to connect the upper and the lower pouches of the esophagus, has also been described in the litterature.

Altorjay et al.[157] described a jejunal interposition in association with a gastric transposition to prevent gastroesophageal reflux in a 5-year old male with esophageal stenosis after repair of atresia at birth. The authors used a 25 cm segment of jejunum interposed between the antrum of the gastric substitution and the duodenum and were successful in avoiding regurgitation.

A review by Liu et al. [158] in which 23 studies were included with a total of 593 patients in whom 45 patients underwent a jejunal interposition. The incidence of anastomotic leak was more significant in this group of patients compared to other esophageal replacement techniques.

However, long term outcomes of jejunal interposition were reported to be positive with a tolerance to oral feeding according to Bax [159]. (Figure 53)

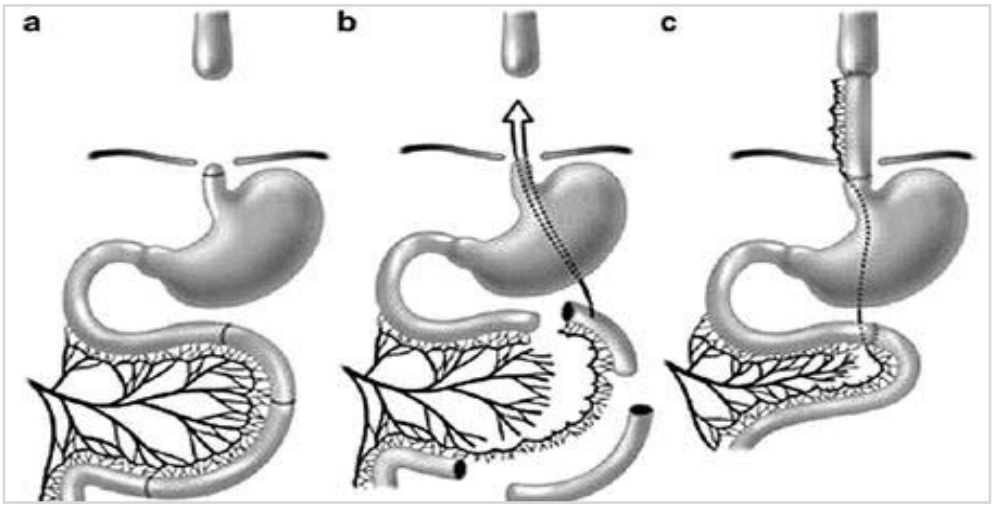


Figure 53[39] : Jejunal interposition.

(a) Suitable portion of the jejunum is chosen.
(b) While the vasculature is preserved, a portion of the jejunum is removed to provide additional length.
(c) The segment of jejunum is passed into the chest in an anteperistaltic fashion. Jejunum continuity is restored.

No esophageal replacement technique is ideal. For this reason, many pediatric surgeons try to preserve the native esophagus whenever it is possible. It is also for this reason that each surgeon tries to use the technique that his team is most used to in order to avoid any subsequent complications [158]. (Table XIX)

Table XIX [158] : Advantages and disadvantages of every esophageal replacement technique

Esophageal replacement technique	Advantages	Disadvantages
Gastric transposition	- Technically easy procedure - Reliable blood supply - Adequate graft length - Single anastomosis in the neck - Thick muscular to stand infection - Lower short-term morbidity - General good long term outcomes	- Bulk of stomach in chest - High risk of reflux - Delayed gastric emptying - Dumping syndrome - Absent peristasis, drain by gravity - Esophagus distal portion is sacrified
Gastric tube	- Adequate graft length - Good blood suply - Retains tubular shape - Rapid food transit	- Long suture lines, prone to leaks and strictures - Anastomosis at neck withrisk of complications - Complicated technique - Neonatal stomach too small to make a tube - Continued acid production by the tube
Colonic interposition	- Better long term outcomes - Low risk of reflux - Adequate length - Occupies little space in chest	- Precarious blood supply - More frequent GER - Likely to become redundant - Need for 3 anastomoses
Jejunal interposition	- Enough length - Peristaltic activity, normal transit - Similar diameters as the esophagus, occupies less space	- Precarious blood supply - Limited length - Technically demanding - Need for 3 anastomoses - More commonn anastomotic complications

e. Thoracoscopic repair :

The benefits of minimally invasive surgery over thoracotomy for reducing pain, scars and long term musculoskeletal deformities including scoliosis have been well documented [160], [161]. The first thoracoscopic repair of a LGEA was carried out in 1999 by Lobe et al. [162]. Since

then, a number of authors have validated the thoracoscopic technique to be equivalent if not superior to open surgery [129], [163], [164].

The patient is positioned semi prone with the right side elevated by 30 degrees. Standard endotracheal intubation is used with no need for endobronchial isolation. Three 3 mm ports are used (Figure 54), although some surgeons use a 5 mm port to facilitate use of an endoclip. A pneumothorax of 3–5 mmHg facilitates lung collapse and visualisation, and a fourth port may be used should lung retraction become necessary. Port placement is crucial due to the limited working space afforded by the neonatal thorax [114].

Bogusz et al. [164] have described a new therapeutic management in 4 patients with LGEA, involving staged thoracoscopic procedure with internal traction without gastrostomy : internal traction was placed between 2 and 6 days of life (Figure 55). A successful repair of native esophagus by the second approach in neonatal period without gastrostomy was achieved in 3 out of 4 neonates, 5–8 days after internal traction placement (Figure 56). No anastomotic leakage was observed, two anastomoses developed stricture and managed with three and four sessions of repeated dilatation. Nasogastric tube feeding started between 5 and 7 days with full oral feeding achieved between 10 and 35 days after anastomosis.

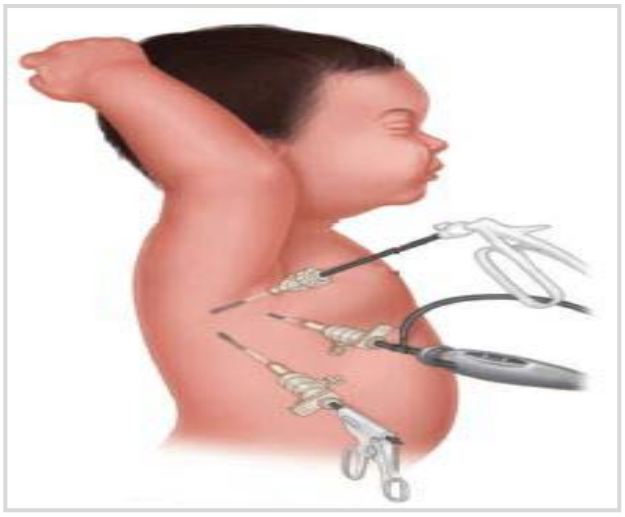


Figure 54[88] : Three ports are routinely for the dissection and intracorporeal anastomosis : 3mm camera port in the 6th intercostal space, 5mm working port in the 3rd intercostal space, and 3mm working port in the 9th or the 10th intercostal space.

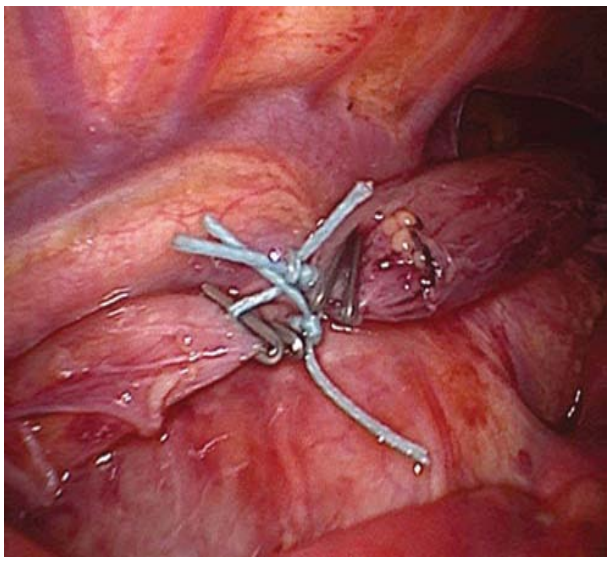


Figure 55[164] : Thoracoscopic view of internal traction of blind esophageal ends.

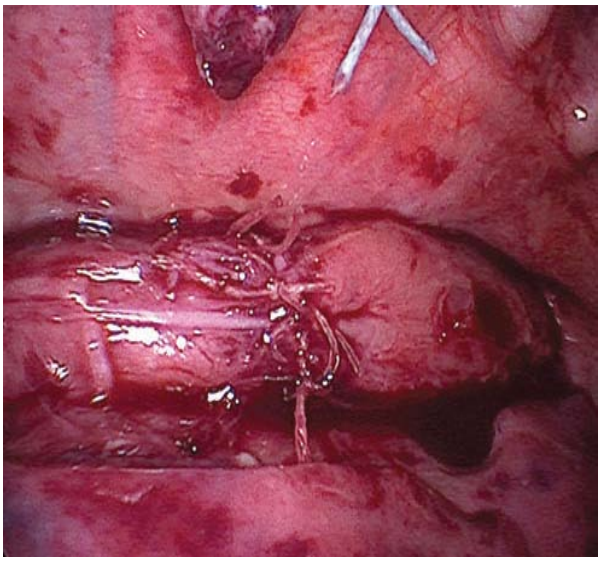


Figure 56[164] : Thoracoscopic view of delayed primary anastomosis.

In our series, we noticed a delay in performing the definitve repair of LGEA. The average waiting time after the initial gastrostomy is 13 months. This is due to the limited number of surgeons qualified to perform this type of surgery in our region, which covers a large population.

The most used surgical option in our series for the definitive repair of LGEA was DPA, which is consistent with the other series listed in Table XX.

Table XX : Comparison of patients number for every surgical option in different series

	DPA	Gastric transposition	Gastric tube	Colonic interposition	Jejunal interposition
Our series	3	2	1	2	-
Series of Ein et al. [149]	8	1	2	-	-
Series of Jensen et al. [1]	25	2	6	-	-
Series of hunter et al. [19]	10	2	3	9	-

3. Postoperative management :

Postoperatively the baby is admitted in neonatal intensive care unit for respiratory monitoring and pain control; transportation shall be done in a portable incubator in order to minimize the heat dissipation [104].

Special care should be directed toward preventing aspiration with frequent oropharyngeal suctioning and elevation of the head of the bed 30-45°. Chest X-ray is carried out to evaluate the pulmonary and the pleural cavity status, the aspect of the mediastinum, the position of the chest tube and of the NG tube.

The duration of postoperative ventilation depends on many factors such as patient weight and prematurity, any associated abnormalities, duration of surgery, tension of the anastomosis, ventilation difficulties during surgery and pain control. A full term newborn with an effective pain control can be extubated early after surgery. On the other hand, a premature baby, or a baby with associated diseases, or when the anesthetist experienced difficulties in keeping the patient well oxygenated during surgery, or when surgery was demanding, all these patients will benefit from a period of postoperative mechanical ventilation [104]. If the primary anastomosis is under tension, a period of 5 days of ventilation, with paralysis and maintenance of the neckflexion, is effective in reducing the incidence of anastomotic leaks, without major side effects [165].

Less invasive forms of respiratory support, such as continuous positive airway pressure (CPAP) and high flow nasal prong oxygen, are being increasing utilised in the neonatal intensive care units. In some units, there has been avoidance of CPAP for infants post EA repair, based on a clinical suspicion that increased upper airway distending pressure may put unwanted pressure on the esophageal anastomosis, compromising repair. However this has neither been tested, nor borne out in the observational literature. In the very preterm patients, well conducted randomised controlled trials have demonstrated that high flow nasal cannulae are as effective as CPAP post-extubation in containing morbidity such as apnea or need for re-intubation [166].

Enteral feedings are begun following an open gastrostomy once the ileus has resolved, and on the day after the operation for the minimally invasive procedures avoiding long term total parenteral nutrition with the associated risks of liver damage. However suck-swallow reflexes and the innate desire to feed may become depressed, making establishment of enteral suck feeding due to oral aversion difficult following a later LGEA repair [167]. For this reason, some authors advocate administration of regular sham-feeds, allowing the neonate to suck from the breast or bottle, with milk collected from the upper pouch via a Replogle tube, and then re-fed back to the neonate via the gastrostomy, facilitating an experience of the sense of satiety (Figure 57). This process requires close supervision by experienced nursing staff, and should not be embarked upon without adequate experience or supervision [166].

In regards to patients who underwent definitive repair of LGEA, enteral feeds may be started gradually via a trans-anastomotic tube on day 1 or 2 postoperatively. During feedings, all patients should be flat with the head of the bed elevated and monitored closely because of the high prevalence of gastroesophageal reflux. Oral feedings are then initiated only after a normal barium swallow conducted 5-7 days after the operation [88].

Regular pharyngeal suction may be useful in the early days after surgery; it is important to clearly mark the length of the suction catheter to prevent its passage beyond the anastomosis with possible damage [104].

The drain in the chest should be maintained until a barium swallow demonstrates the integrity of the esophageal anastomosis. A fistula may be identified directly on the barium swallow or may be inferred from the appearance of saliva in the chest tube. Alternately, an extrapleural effusion on chest radiographs may be associated with anastomotic leakage. In order to protect the anastomosis, the chest tube should remain if a significant fistula appears. The vast majority of esophageal leaks controlled by chest tube drainage should spontaneously seal with time [88].

Antibiotic therapy, either initiated preoperatively or administered at induction, will be continued until the chest tubes are removed.

Proton pomp inhibitors are routinely administered to at least the age of 12 months [107].

The patient is discharged when feeding well and gaining weight. Parents should be warned about the signs of complications such as gastroesophageal reflux, tracheomalacia, anastomotic stricture, and recurrent fistula [4].

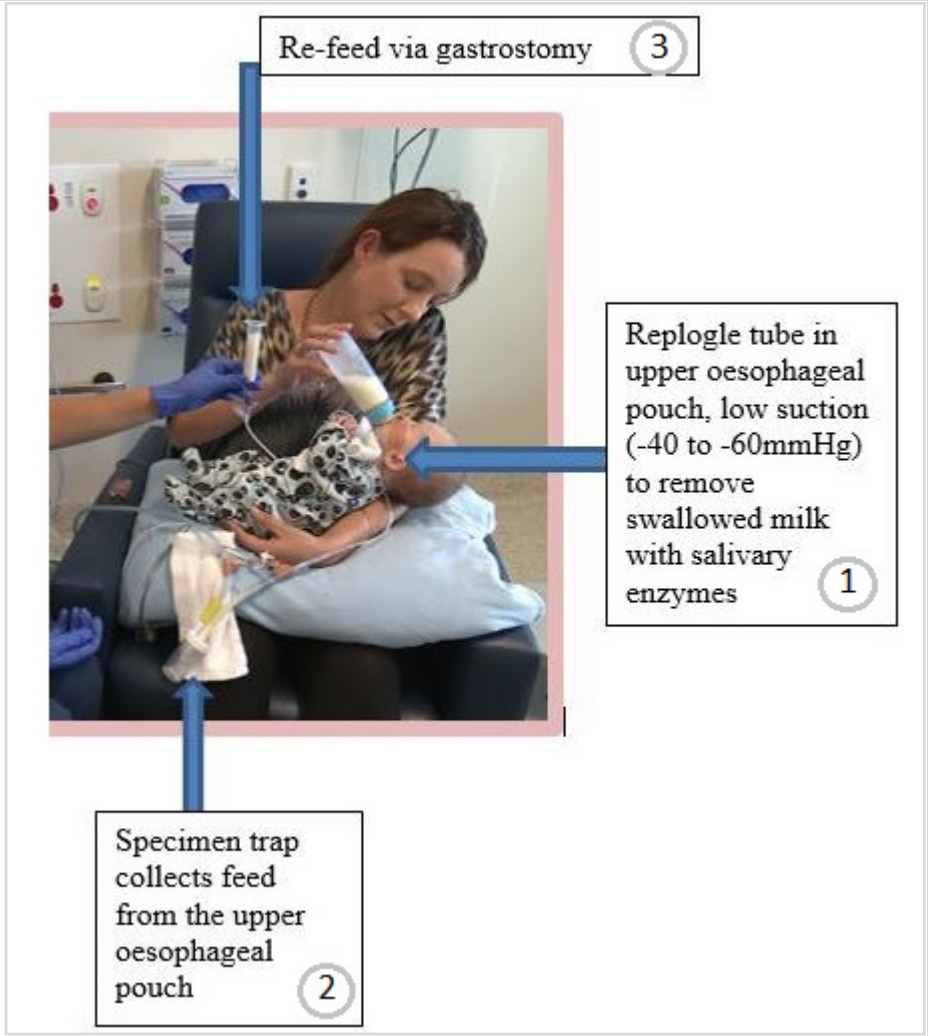


Figure 57[168] : Sham feeding steps

VII. Postoperative complications :

Complications after LGEA repair can be divided into short term (<30 days) and long term complications (>30 days).

1. Short-term complications :

The most common early complications after LGEA repair are anastomotic stricture and anastomotic leakage [148], [158]. These complications can be life-threatening, hence the need for careful monitoring in intensive care unit.

Table XXI summarizes the main early complications found in our series and in the literature.

Table XXI : Comparison of the incidence of early postoperative complications

Series	Cases number	Anastomotic leakage	Anastomotic stricture	Fistula recurrence	Pneumonia
Our series	21	3 (14%)	-	1 (4%)	5 (24%)
Series of hunter el al. [19]	28	4 (14%)	8 (28%)	-	1 (3%)
Series of Galo et al. [18]	24	11 (46%)	7 (29%)	-	6 (25%)
Series of Al-Shanafey [13]	17	6 (35%)	2 (12%)	-	1 (6%)
Series of Zani et al. [169]	12	7 (58%)	7 (58%)	-	2 (17%)

Comparing our series with the others, we notice :

- A high incidence of pneumonia of 24%, comparable with that of Galo et al. series.
- Low rate of anastomotic leakage in our series, the highest rate was recorded in the series of Zani et al.
- No case of anastomotic stricture was recorded in our series, while the rate of anastomotic stenosis ranges from 12% to 58% in the other series.

1.1. Anastomotic stricture :

Anastomotic stricture is the most common early complication after a LGEA repair and it has been also recorded in the late postoperative times [158]. Its incidence varies from 8% to 62,2% depending on the surgical option performed and on the criteria used to define a stricture [148].

Esophageal stricture can be asymptomatic or cause symptoms such as hypersialorrhea, prolonged feeding, incomplete feeding, coughing. Stricture is confirmed then by a contrast study showing a narrowing of >50% of the lumen [170]. (Figure 58)

Parolini et al. [171] have conducted a retrospective longitudinal study in which 35 patients with EA were included, with routine endoscopy performed 1 month after repair. The aim of the study was to identify identify risk factors for predicting the development of anastomotic stricture following EA repair. The authors have found GER, anastomotic tension as well as LGEA had an increased risk of developing an anastomotic stricture.

Clinically significant narrowing at the site of the esophageal anastomosis is treated by dilation performed via bougienage or balloon dilators. The radial dilating forces generated during balloon dilatation are considered to be less traumatic than the longitudinal shearing forces caused by conventional bougienage [172]. Balloon dilatation is performed under fluoroscopic control by passing a guide wire through the stricture, over which a balloon dilator of appropriate size is introduced. Its position is confirmed by partially filling the balloon with contrast medium, so that “wasting” due to the stricture is centrally located (Figure 59). The balloon is then maximally inflated using dilute contrast medium to dilate the stricture. A contrast esophagogram is performed after removing the balloon to ensure that there has been no injury perforation.

Topical application of mitomycin C, intralesional steroids and placement of stent are viable options in patients with recurrent strictures (defined by 3 anastomotic structure relapses requiring dilation) [107].

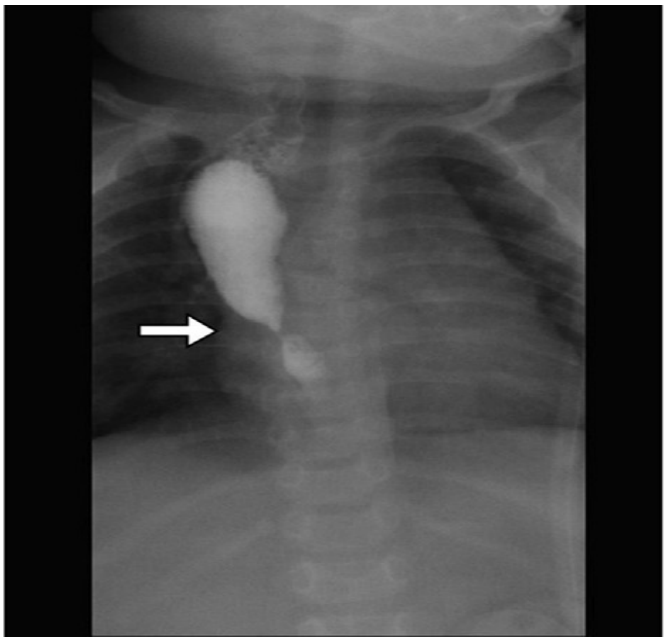


Figure 58 [173] : Contrast study shows a Medium and focal narrowing (arrow) at the mid-to-lower thoracic level with proximal dilation.

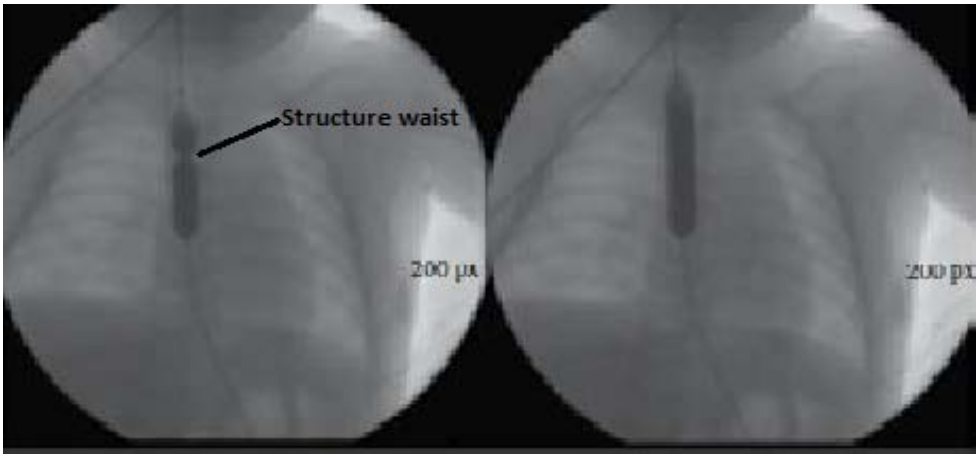


Figure 59 [14] : Balloon dilation of an esophageal anastomotic stricture. 'Wasting' appearance shows the site of structure.

1.2. Anastomotic leakage :

Anastomotic leakage occurs in 1 of 5 patients after LGEA repair [148] and classically happens on the 3rd or 4th postoperative day; many are relatively asymptomatic and therefore go clinically unnoticed. The clinical manifestations depend upon the size of the leak [88]. Leakage is usually manifested by pneumothorax and salivary drainage from the chest drain. The only

evidence of a tiny asymptomatic leak may be the radiological finding of a bit of extravasated water-soluble contrast at the sight of the anastomosis, usually when the first routine contrast swallow is done on the 5th or the 7th postoperative day (Figure 60).

Provided that a transanastomotic tube is in place, it is usually possible to control the leak with an adequately sized chest drain. With adequate drainage, broad-spectrum antibiotics, and total parenteral nutrition, the esophagus will usually heal, although a prolonged period with a chest tube may be necessary [174]. Some surgeons have used hyoscine patches in an attempt to "dry up" the salivary leak. Others advocate early re-exploration (<48 hours), with direct repair of the esophagus if possible, and the establishment of satisfactory drainage where a major leak is suspected [175].

Basuguy et al. [176] described a new conservative approach to manage anastomotic leakage ; the nasogastric catheter was fluoroscopically converted into a nasojejunal catheter using a guidewire and feeding continued (Figure 63). The results were overall promising.

When conservative management fails with uncontrolled sepsis, the establishment of a cervical esophagostomy and a feeding gastrostomy are essential[14].

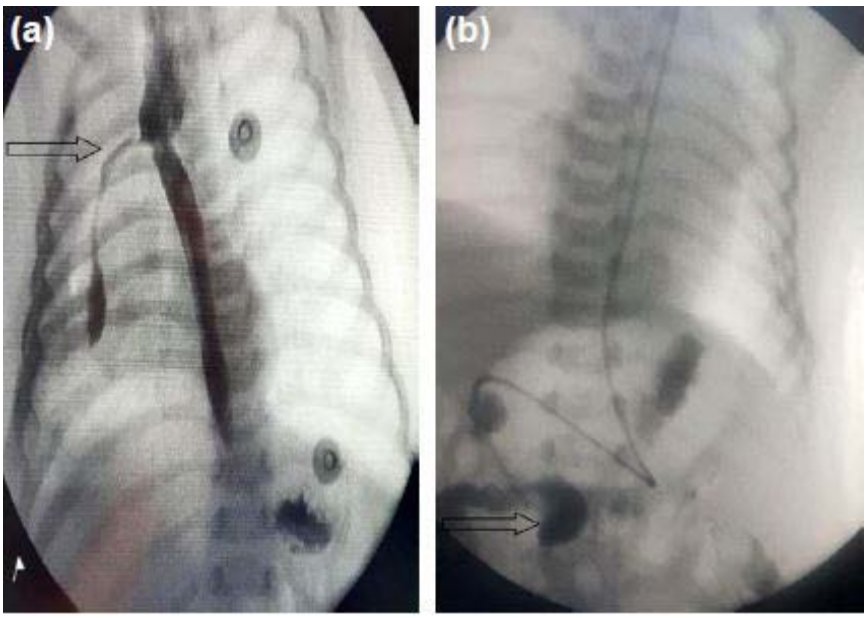


Figure 60 [176] : (**a**) Anastomotic leakage. (**b**) Nasojejunal catheter

1.3. Fistula recurrence :

Recurrent TEF occurs in 3% to 15% of patients after initial ligation [177]. Fistula recurrence has been attributed to anastomotic leak with local inflammation and erosion through the previous site of TEF repair, but the possibility of a missed upper pouch fistula should always be considered. Although recurrent TEF typically occurs in the early postoperative period, it may not be recognized for months to years. Symptoms include recurrent chest infections, coughing and choking attacks during feeding. These symptoms are often difficult to distinguish from those of tracheomalacia and GER that occur frequently in infants born with EA [177].

The diagnosis may be suggested by an air-filled esophagus on plain radiographs of the chest. Routine contrast swallow studies will miss up to 50% of recurrent fistulas [87]. The investigation of choice remains a combined esophagoscopy and bronchoscopy [14] ; rigid bronchoscopy is performed initially, and the site of the original fistula is carefully examined. The fistula is gently probed with a ureteric catheter, and methylene blue is carefully instilled into the fistula pit. Synchronous flexible esophagoscopy is performed to see if blue dye can be seen entering the esophagus. Should this fail to demonstrate the fistula, an “air/water test” is a useful supplementary investigation. The esophagus is filled with water and positive pressure ventilation applied to the bronchoscope. Occasionally, bubbles of air can be seen emanating from the fistula’s opening intothe esophagus [14]. (Figure 61)

Several strategies have been described to manage the recurrent TEF. The traditional approach is formal repair via right thoracotomy. Early open surgical closure of the fistula is not easy, due to the local inflammatory process and a reduced tissue quality, and it is associated with significant morbidity and failure rate [178]. Some authors advocate the use of vascularized pleura, pericardium or intercostal muscleflaps for separation of the suture lines in order to prevent second recurrences [177], [179].

This high rate of postopen surgery complications has led many authors to advocate endoscopic management as the first line of treatment of recurrent TEF. Endoscopic techniques fall into three general categories: obstruction of the fistula tract, de-epithelialization of the

fistula, or a combination of the two. Tissue adhesives are used primarily to obstruct the fistula tract. Removal of the fistula tracts epithelium can be accomplished with electrocautery, mechanical abrasion, sclerosants, and lasers [180]. No specific endoscopic treatments have been demonstrated to be superior to others [178]. (Figure 62)

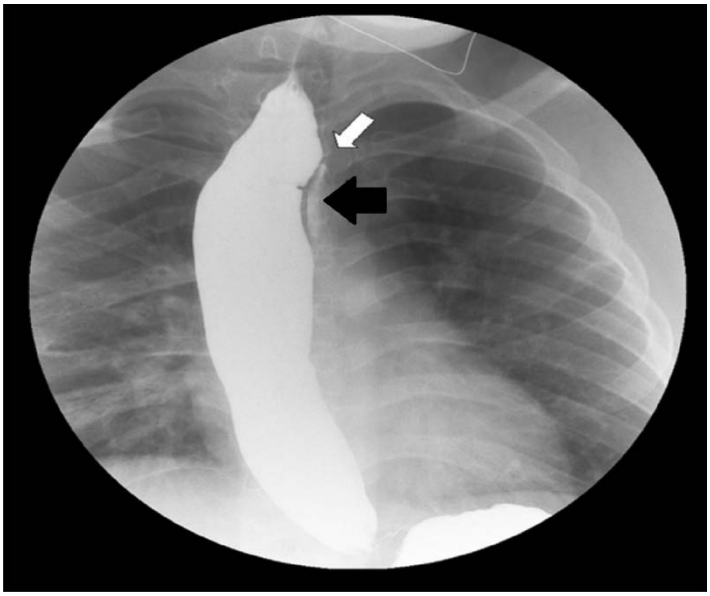


Figure 61 [181] : Barium esophagogram demonstrating fistulous connection (white arrow) with contrast drainage into the trachea (black arrow).

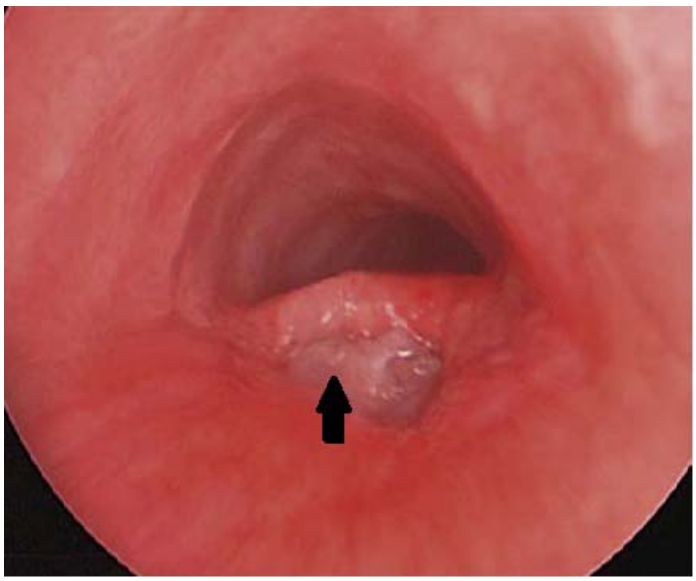


Figure 62 [39] : Bronchoscopic view of a successful recurrent TEF closure with a surgisis (black arrow).

1.4. Vocal cord dysfunction :

Another complication that is becoming increasingly recognized is injury to the recurrent laryngeal nerves during EA repair. In a retrospective review of 150 patients undergoing repair of EA and associated anomalies, five patients (3%) were found to have vocal fold paralysis on subsequent evaluation. Bilateral paralysis was found in three patients and two patients had unilateral paralysis. In this study, the etiology of the paralysis was difficult to assess. However, the authors recommend preoperative laryngoscopy or bronchoscopy to identify infants with congenital vocal cord paralysis prior to operative repair. This is especially true for patients requiring revision surgery [182].

An older review found a 12% incidence of vocal cord injury following EA repair [183].Recent study by Conforti et al. [184] have described vocal cord paralysis in 36% in patients with pure EA and the H-type TEF.

2. Long term complications :

2.1. Gastroesophageal reflux (GER) :

Gastroesophageal reflux (GER) remains a frequent long-term morbidity in patients undergoing EA repair with a reported rate ranging from 17% to 75% [185]-[187]. The causes of GER are multifactorial and include developmental neuronal dysfunction in the lower esophagus, effects of the surgical mobilization of the lower pouch, and the vagal innervation as well as disturbance of the esophagogastric junction and the Hiss angle.

The diagnosis of pathologic GER is suspected in patients with symptoms of vomiting, dysphagia, and recurrent anastomotic stenosis, which is occasionally associated with the impaction of a foreign body or food bolus. In addition, respiratory symptoms such as stridor, cyanotic spells, recurrent pneumonia, and reactive airways disease may indicate GER rather than other conditions such as tracheomalacia.

24 hours pH monitoring remains the gold standard investigation to explore acid GER [188]. Esophageal impedance monitoring allows the diagnosis of all associated non-acidic GER

episodes [189] . It has recently been reported that reflux episodes are more frequent and longer in children with EA compared to control children, and that the majority of reflux episodes in EA are non-acidic [189].

In infants and children with pathologic GER, aggressive medical management typically consists of thickening of feedings, positioning of the infant in a prone or upright posture, and administration of acid reduction agents such as histamine-2 blockers, proton pump inhibitors, and prokinetic agents. However antireflux medication is successful in only half of the cases[190]. Failure to control GER with full medical therapy is an indication for fundoplication. Fundoplication rates following surgery for EA vary widely between surgery centers (6%–45%), reflecting the varied enthusiasm for antireflux operations in the clinical setting of GER [191], [192]. There are several reasons for caution when considering fundoplication in EA patients. Dysphagia may be aggravated as a consequence of underlying foregut dysmotility. Fundoplication following EA repair has a higher failure rate (15%–38%) than is generally observed in otherwise normal patients with isolated GER[193]. Some authors recommend a partial wrap (Thal) fundoplication because of the lower incidence of postoperative dysphagia[191]. Despite such concerns, many pediatric surgeons prefer a short (1.5–2.0 cm) 360° floppy Nissen wrap for its proven effectiveness in reducing GER[193].

A concern of long standing GER and esophagitis is that chronic mucosal injury can progress to metaplasia followed by dysplasia and ultimately adenocarcinoma of the esophagus. The prevalence of Barrett's esophagus (BE) after EA repair has been reported to range from 5% to 43% [194]. Schneider et al. [194] conducted a prospective multi-institutional study in which they examined 123 patients with EA and subjected all patients to multistaged endoscopic biopsies under general anesthesia. They found there was nearly a five fold increase rate of BE in patients who had pure EA, suggesting that long gap length,which is more common in pure EA, likely increases the risk of BE. Interestingly, there was no association between patient who reported symptoms of GER and BE, reinforcing the importance of lifelong endoscopic surveillance with biopsies, even in asymptomatic EA patients.

Reconstruction of the esophagus with a gastric tube interposition has been found to be associated with a high incidence of cervical BE [195]. On the other hand, there have been no reported cases of BE in patients undergoing colonic interposition for LGEA [196], [197].

The risk of esophageal cancer in patients after EA repair is unknown and controversial. There have been 9 reported cases of esophageal cancer after EA repair, 7 squamous cell and 2 adenocarcinoma [72,83–88]. All nine underwent primary EA repair with no description of esophageal gap length. The mean age of the patients at the time of esophageal cancer diagnosis was 38 years [185], [198].

The incidence of Barrett's esophagus and esophageal cancer in repaired EA is predicted to increase as more patients are entering older adulthood. Therefore, endoscopic surveillance with multistage biopsies occurring throughout the esophagus including the gastroesophageal junction, anastomotic site and any macroscopic lesions should be performed when EA patients reach their teenage years and then routinely thereafter. Patients found to have Barrett's without dysplasia should undergo repeat endoscopy/multistaged biopsies every 3 years. Those found to have Barrett's with dysplasia should undergo repeat endoscopy/multistaged biopsies every 6 months with consideration for mucosal resection. Patients without Barrett's should undergo surveillance endoscopy/multistaged biopsies every 5–10 years [185].

2.2. Tracheomalacia :

Tracheomalacia is a generalized or localized weakness of the trachea that allows the anterior and posterior walls to come together during expiration or coughing [199]. Tracheomalacia has been noted to affect up to 75% of pathologic specimens from patients suffering from EA; however, the condition is problematic or significant in approximately 10% to 25% of infants after repair of EA, approximately half of whom require surgical correction.

Infants with tracheomalacia demonstrate expiratory stridor, which may result in episodes of desaturation, apnea, cyanosis, and bradycardia (often associated with feeding), and life-threatening so-called “dying episodes.” Severe tracheomalacia may be evident in the early postoperative period, when it proves difficult to wean the infant from the ventilator [14].

Indications of the severity of tracheomalacia include ventilator dependency, respiratory distress characterized by stridor and chronic carbon dioxide retention, and “dying episodes.” Full investigation for severe GER and recurrent TEF is advisable alongside evaluation of tracheomalacia, as aspiration secondary to GER and a recurrent fistula can mimic these symptoms. The extent of tracheomalacia is assessed by bronchoscopy under conditions of spontaneous respiration. The lumen of the trachea is significantly compressed anteroposteriorly and assumes a scabbard-like appearance during expiration due to tracheal cartilage deficiency. A further contribution is often made by the upper esophagus, which may bulge posteriorly into the airway. Tracheobronchomalacia can extend beyond the carina into the main stem bronchi. (Figure 63)

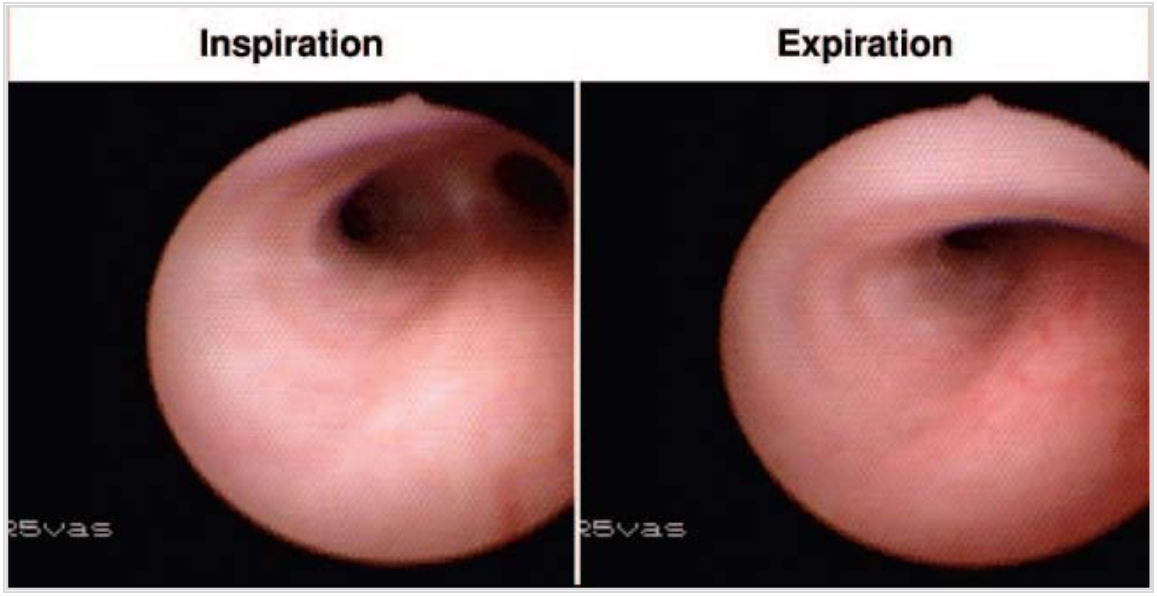


Figure 63[200] : Bronchoscopic findings in tracheomalacia. During inspiration, the trachea remains patent and the carina is easily visible. During expiration, the trachea collapses on itself, obstructing the carina and distal airways.

As tracheomalacia can be self-limiting, surgical intervention is reserved for patients with life-threatening symptoms. Treatment options include CPAP, aortopexy, tracheostomy, and more recently, tracheal stenting [201]. CPAP is a useful temporizing measure but may be required for several weeks.

Aortopexy is traditionally performed by anterior left thoracotomy through the third interspace. The left lobe of the thymus is excised to gain access to the root of the aorta taking care not to damage the phrenic nerve. The sutures are passed through the periosteum of the manubrium and tied, hitching the aorta forward, there by relieving compressive forces on the trachea. Although this operation can not resolve distal tracheobronchomalacia, it often provides immediate dramatic symptomatic relief. Failure of aortopexy may be an indication for tracheostomy, although some authors advocate tracheal stenting in this situation [192], [201].

2.3. Esophageal dymotility :

Esophageal dysmotility is present in the majority of children born with EA and is a key factor in the pathophysiology of numerous EA-associated comorbidities such as dysphagia. 36% to 52% of adults who underwent EA repair experience dysphagia [202], [203].

Several studies, whose purpose was to provide a better understanding of the pathophysiology underlying dysmotility, have examined the esophageal tissue of patients born with EA (obtained during either autopsy or surgical repair) or in animal models [204], [205]. These studies have identified abnormalities in the myenteric plexus of the proximal esophageal segment (such as hypoplasia and abnormal interganglionic connections) [204] and reduced density and immaturity of interstitial cells of Cajal (intestinal cells that are important for gastrointestinal motility) in the proximal and distal segment [205]. One study in two patients with TEF who underwent high resolution manometry (HRM) before surgery showed defects in esophageal peristalsis, providing direct evidence that neural innervation may be aberrant before surgery [206]. By contrast, another study in two patients born with LGEA who underwent manometry recordings of the proximal and distal esophageal segments, demonstrated seemingly normal contractile patterns [207]. That is, the motor patterns in these patients became aberrant only after surgical repair. Indeed, surgical repair may worsen dysmotility by potentially causing denervation of extrinsic inputs to the proximal esophageal muscle and the enteric nervous system of the distal esophagus [208]. However, owing to the extremely small

numbers, no firm conclusions regarding differences in surgical techniques and their influence on esophageal motility can be drawn at this stage.

2.4. Colonic interposition grafts redundancy :

A reported late complication of colonic interposition is dilation and tortuosity of the transposed colon reported in 3% to 12% of patients [209]. Over time, it is believed that the lack of motility as well as the negative intrathoracic pressure in the chest results in redundancy of the colon. Retrosternal placement of the colon conduit has been reported to result in a lower incidence of graft redundancy [196].

Hamza et al. [196] reported on 775 patients undergoing colonic interposition of which 17 (2%) developed redundancy of the conduit with 5 requiring revision. Another contemporary study of 115 children undergoing esophagocoloplasty reported no redundancy of the colongraft utilizing either the retrosternal or posterior mediastinal placement [210]. There are currently no clinical guidelines/recommendations on whether routine surveillance fluoroscopy should be performed in patients undergoing colonic interposition or if it should only be performed in symptomatic patients.

2.5. Nutrition and growth outcome :

Although recent advances in surgical techniques, neonatal intensive care, and nutritional support have led to a significant decrease of malnutrition and stunting growth , the latter remains present in nearly one-third of children at age 5 years [186].

In a study by Gallo et al. [18] comparing long term outcomes in patients undergoing gastric transposition and jejunal interposition for LGEA, poor growth was found in both cohorts of patients at 8–14 years after esophageal replacement. 44% of gastric transposition patients and 26% of jejunal interposition patients had weight/age anthropometric measurements that were two standard deviations below average. Another study of patients with LGEA compared those repaired with a gastric tube to those undergoing a colon interposition. There was no difference

in nutrition or growth between the two techniques but on long term follow-up, nearly all patients were at or below the 10th percentile for height and weight [211].

Therefore, nutrition and growth monitoring is essential, especially at the time of growth peaks (first 4 years of life, puberty) [186].

2.6. Skeletal abnormalities :

Skeletal abnormalities including scoliosis, winged scapula and anterior chest wall deformities are common in published series of EA patients. Contributing factors include intrinsic vertebral anomalies common in EA patients as well as sequelae of surgical correction of EA via thoracotomy.

Clinically significant scoliosis has been reported in 56% of patients after open EA repair which represents a 13 fold increase as compared to the general population [212]. In a majority of patients, the scoliosis did not require surgical correction and was followed clinically. Winged scapula has been reported in 24% and anterior chestwall deformities (including pectus carinatum and excavatum), and in 20% after open EA repair [212], [213].

Moreover, chronic pain after thoracic surgery is a serious problem, at least in adults, and has been reported in >50% of patients [214]. These negative consequences of a thoracotomy can be alleviated by using the thoracoscopic approach.

2.7. Long term quality of life :

Survival imporvement of LGEA patients born in the modern era has prompted detailed analysis of morbidity with emphasis on long-term outcome.

Koivusalo et al. [215] examined various disease specific quality of life (QOL) outcomes as well as overall health-related quality of life (HRQOL) using validated questionnaires. One hundred twenty eight adults (median age: 38 years) who underwent EA repair as infants were compared to 162 healthy control subjects (median age :36 years). The EA cohort included patients undergoing primary repair as well as esophageal replacement with colon interpositions grafts and gastric tube reconstructions. Results of the 36 item Respiratory Symptoms-Related

Quality of Life Index (RSRQLI) demonstrated increased respiratory symptoms with increased severity in EA patients as compared to healthy respondents (7% vs. 1.8%, respectively,). Gastrointestinal Quality of Life Index (GIQLI) was equivalent between EA repair patients and healthy controls. Similarly, there was no difference in psychosocial functioning, assessed by RosenbergSelf-esteem Scale, Beck Depression Index and Cohen's Test for Life Management Ability between groups. Therewas no overall difference in HRQOL utilizing both tools between EA repairpatients and healthy controls.

Ludman and Spitz [216] examined medium term QOL (mean of 12.99 years) utilizing a modified GIQLI in 28 LGEA who underwent gastric transposition. They reported unimpaired QOL after primary gastric transposition.

Dingemann et al. [217] reported on long term HRQOL in 90 patients with complex EA, 27 of which underwent esophageal replacement. At a mean of 14.5 years, there was no difference in HRQOL between complex EA patients and healthy controls as measuredby KIDSCREEN27 which assesses subjective health and well-being inchildren and adolescents. However, the subgroup of patients who underwent esophageal replacement reported poorer GIQLI scores and physical well-being scores as compared to healthy controls.

VIII. Prognosis :

1. Prognostic classifications :

1.1. Waterston classification :

Waterston et al have established in 1962 the first prognostic classification for infants with EA, in which they have stratified patients into three categories according to birth weight, the presence of pneumonia and the identification of other congenital anomalies [21]. (Table XXII)

Although the Waterston classification continues to be used to compare results between centers, many authors have questioned its current validity in caring for these infants.

Table XXII : Waterston classification [21]

Category	Definition	Surgical timing
A	Weight > 2500g with no pneumonia or other congenital anomalies	Immediate surgery
B	1800g<weight<2500g or mild pneumonia or moderate anomalies	Short-term delay, plus stablisitation
C	Weight< 1800g or severe pneumonia or severe anomalies	Staged repair

1.2. Montreal classification :

With advances in neonatal critical care, more low-birth-weight infants are surviving and more treatment options are available for infants with multiple congenital anomalies. As a result, a search for modern criteria for survival has produced several new classification schemes.

In 1993, Poenaru et al. established a new risk classification. They suggested that only severe pulmonary dysfunction with a preoperative mechanical ventilation requirement and severe associated anomalies were independent predictors of survival [25]. (Table XXIII)

Table XXIII : Montreal classification [25]

Classes	Definition
Class I	No ventilator dependence nor minor or major anomalies, or ventilator dependence and no minor or major anomalies.
Class II	Ventilator dependence and major anomalies, or no ventilator dependence and life-threating anomalies.

1.3. Spitz classification :

In 1994 , Spitz and colleagues [22] provided a new risk grading system based on birthweight and presence or absence of congenital heart disease, that is now widely accepted as applicable to the modern era. (Table XXIV)

Table XXIV : Spitz classification [22]

Groups	Definition	Survival rate
Group I	Weight > 1500g and no major cardiac anomaly	98%
Group II	Weight < 1500g or major cardiac anomaly	82%
Group III	Weight > 1500g and major cardiac anomaly	50%

A major cardiac anomaly is defined as either cyanotic congenital heart disease requiring palliative or curative surgery, or non-cyanogenic heart disease requiring medical or surgical treatment for heart failure. The progress made in the management of newborns with low birth weight has made it possible to relativize the pejorative value of this criterion, compared to other associated malformations, which is currently, for all authors, the essential prognostic factor and the main cause of death in esophageal atresias.

1.4. Yamoto classification :

In 2018, Yamoto el al. [218] proposed a new prognostic classification close to the Spitz classification using the presence of cardiac anomalies and birth weight. (Table XXV)

Table XXV : Yamoto classification [218]

Complex cardiac anomaly	Weight	Risk classification
Absent	>2000g 1000g-2000g <1000g	Class I (low risk) Class II (intermediate risk) Class III (high risk)
Present	>2000g 1000g-2000g <1000g	Class II (intermediate risk) Class III (high risk) Class IV (super high risk)

Unlike the previous classifications that have concentrated on associated medical conditions, Brown and Tam proposed aclassification based on gap lengths to define the magnitude of the surgical problems in EA and correlate them with outcome. Infants with LGEA experience a high morbidity from anastomotic leak, stricture,failure/redo operations, and esophageal replacement [219].

2. Mortality :

The survival rate of patients with LGEA has improved significantly from 35% [220] to 95% [16], [18] and reaches 100% in some series.

In our series, despite the marked improvement, but the survival rate (57%) is still lower than that in foreign series (Table XXVI) ;5 patients passed away after undergoing the initial gastrostomy (24%), 1 patient after delayed primary anastomosis, 1 patient after reverse gastric tube and 1 after colonic interposition.

Table XXVI : Comparison of survival rate in different series

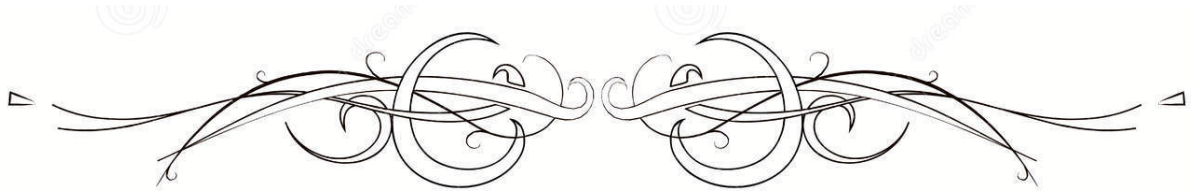
Series	Number of patients	Survival rate
Our series	21	57%
Series of Bagolan et al. [41]	57	81%
Series of Maheshwari et al. [167]	15	87%
Series of Hunter el al. [19]	28	93%
Series of Powell et al. [221]	44	84%

IX. Recommendations :

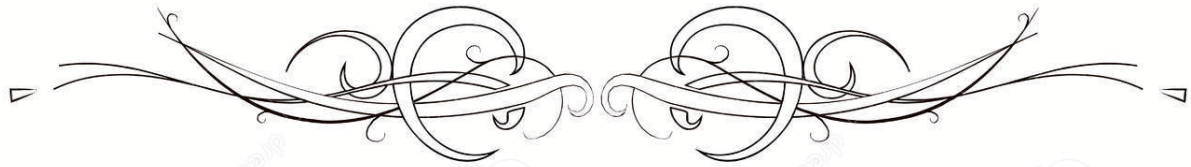
In order to improve the prognosis of LGEA in our context, it is necessary :

- Follow regularly the pregnancies up with at least 3 obstetrical ultrasounds and search for suggestive sonographic signs of the diagnosis.
- Raise awarness among health care professionals about the need to include the check of esophageal continuity as part of the routine examination of all newborns in the delivery room.
- Start without a delay an initial management including :
  - Placing patients in semi setting position.
  - Continous suction of the upper pouch.
  - Prohibit any oral feeding.
  - Insertion of a peripheral veinous catheter.

- In patients with type A LGEA, a gastrostomy tube placement must be performed in first days of life. A second look must be done, at the latest after 12 weeks, to assess the remaining gap. If the remaining gap is less than or equal to 2 cm, a myotomy should be performed allowing to make an end-to-end anastomosis and preserve the native esophagus. If the remaining gap is greater than 2 cm, an esophageal replacement is needed.
- In patients with type C LGEA, ligation of TEF must be carried out in early stage of life. In the meantime Foker process is recommended to stimulate esophageal growth and reduce the waiting time before the definitive repair of LGEA. The first stage must be completed by a gastrostomy tube placement to allow enteral feeding. After 8 to 12 weeks, a second look, with complete dissection of the upper pouch, must be done to evaluate the remaining gap. If the remaining gap is less than or equal to 2 cm, myotomy with an end-to-end anastomosis should be performed. If the gap is greater than 2 cm, then an esophageal subsitution is required.
- Respect of asepsis rules and implement stringent measures to prevent nosocomial infections.
- Manage adequately the early post operative complications.
- Screen and treat associated malformations.
- Explain to parents how to manage the gastrostomy tube at home and warn them about the signs of complications such as gastroesophageal reflux, tracheomalacia, anastomotic stricture, and recurrent fistula
- Set up an interdisciplinary follow-up program including surgeons, gastroenterologists, pulmonologists, otolaryngologists, nutrition counseling and others, with one specialist leading.
- Establish a structured education program for adolescents with LGEA to facilitate their transition from pediatric to adult care.
- Create a national registry for EA in collaboration with all hospitals treating this malformation in Morocco in order to: assess the prevalence of EA and especially LGEA, improve its management and ensure a short and long term follow-up.



CONCLUSION



Long gap esophageal atresia is a rare variant of esophageal atresia. Certainly its prognosis has improved, but its management remains a major challenge for pediatric surgeons and affected patients. What contributes to this situation, is the lack of consensus regarding the definition of LGEA, gap measurement techniques and the ideal surgical option. Moreover, LGEA is associated with a high rate of complications, therefore, a close monitoring both in the short and long terme is essential.

At the end of our study, we note :

- A low rate of prenatal diagnosis.
- Routine screening for EA is not performed in all newborns in the delivery room.
- A delay in the definitve repair of LGEA
- All of the above factors, in addition to the other malformations found in this group of patients, have contributed to the fact that the mortality rate is still high compared to developed countries.

In conclusion, our diagnostic and therapeutic management requires additional efforts to improve survival and offer patients operated for LGEA a better quality of life.

ANNEX

Annex

Operating sheet

I. Patient identity :

Name :
File number :
Gender : ◯M ◯F
Entry age : ……Hours or ……Days
Admission date :
Discharge date :

II. Anamnesis :

1- Cause of hospitalization : .....

2- Maternal medical history :

- Age : .....
- Gravidity : .....
- Parity : .....
- Maternal diseases : .....
- Term of the pregnancy (weeks of amenorrhea) : .....
- Delivery mode :
  ◯Vaginal delivery ◯Caesarean section
- Familial medical history of esophageal atresia : ◯Yes ◯No
- Pregnancy follow up : ◯Yes ◯No
- Oral feeding attempt : ◯Yes ◯No

3- History of the illness :

A- Prenatal diagnosis : ◯Yes ◯No

B- Functional signs :

- Cyanosis : ◯Yes ◯No
- Cough : ◯Yes ◯No
- Vomiting : ◯Yes ◯No
- Hypersialorrhea : ◯Yes ◯No
- Fever : ◯Yes ◯No
- Other : .....

III. Physical examination :

-Birth weight (grammes) : .....
-Apgar score : 1st min:.....
5th min:.....
10th min:.....
- NG tube insertion test : ◯Positive ◯Negative
- Other examination anomalies : .....

IV. Paraclinical examination :

-Chest x-ray :.....
-Pelvic and abdominal ultrasound :.....
-Echocardiography :.....
-Associated malformations : § Vertebral :
§ Anorectal :
§ Cardiovascular :
§ Tracheal :
§ Renal :
§ Limbs :
-Other :.....

V. Management :

- Duration of hospitalization in intensive care before surgery : .....
- Patient operated : ◯Yes ◯No
- Surgical approach : ◯Right thoracotomy ◯Left thoracotomy
◯Laparotomy ◯Cervical approach ◯Thoracoscopy
- Delayed primary repair : ◯Gastrostomy ◯Esophagostomy
- Esophageal replacement : ◯Reverse gastric tube ◯Gastric pull up
◯Colonic interposition ◯ Jejunal interposition
- Duration of hospitalization in intensive care after surgery : .....

VI. Complications :

1. Short term complications :

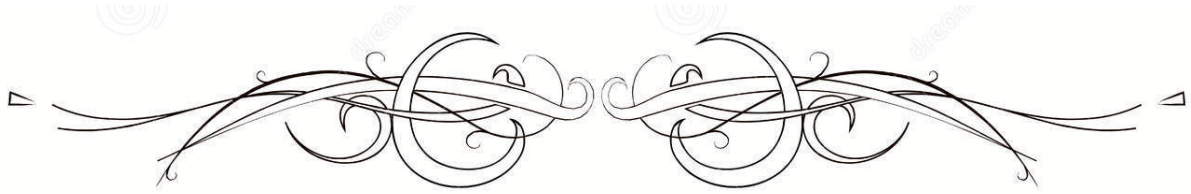
-◯Anastomotic leak : -◯Anastomotic stricture :
-Respiratory complications : ◯Pneumonia ◯PNO ◯Pleural effusion :
-◯Dehydration : -◯Septic shock :
-Other : .....

2. Long term complications :

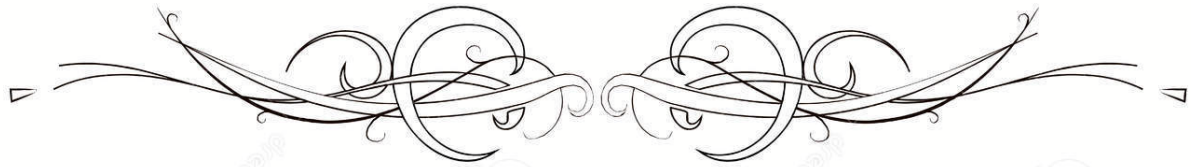
-◯Normal oral feeding :◯Yes ◯No -◯Growth delay : -◯Dysphagia :
-◯Gastroesophageal reflux : -◯Orthopedic deformities :

3. Mortality :

- Death : ◯ Yes ◯No - Cause of death :..... Survival time : .....



ABSTRACT



Abstract

Long gap esophageal atresia occurs in 5% to 10% in all esophageal atresia cases. It is both a medical and surgical emergency that must be diagnosed at the latest in the delivery room. Its management includes initially the pediatric intensivists and pediatric surgeons, and in the long term gastroenterologists, pulmonologists, otolaryngologists, nutrition counselors. Several surgical techniques have been described to manage long gap esophageal atresia without any of them proving to be better.

Our study, which took place in the department of pediatric general surgery of the University Hospital Center Mohamed 6 of Marrakesh, included 21 patients over a period of 7 years from January 2013 to December 2020.

Mean birth weight was 2790g and prematurity rate was 19%. Associated malformations rate was 38%; cardiovascular defects were the most common (19%). 13 patients had a Gross type A EA (62%) and 8 had a Gross type C (38%). All patients underwent an initial gastrostomy, 8 of them underwent later a definitve repair of LGEA; 3 underwent a delayed primary anastomosis, 2 a colonic interposition, 2 a gastric transposition and 1 a reverse gastric tube. Complications were reported in 67% of patients and the mortality rate was 43%.

In the light of this study, we have shown that the survival rate is still pejorative in our context compared to developed countries. This can only incite us to make more efforts and to correct the deficiencies found in our management in order to offer a better quality of life to our patients.

Résumé

L'atrésie de l'œsophage grand écart représente 5 à 10% de tous les cas d'atrésie de l'œsophage. C'est une urgence à la fois médicale et chirurgicale dont le diagnostic doit être fait au plus tard à la salle de naissance. Sa prise en charge nécessite dans un premier temps l'intervention des réanimateurs pédiatres et les chirurgiens pédiatres, et à long terme celle des gastrologues, des pneumologues, des oto-rhino-laryngologistes et des diététiciens. Plusieurs techniques chirurgicales ont été décrites pour la prise en charge de l'atrésie de l'œsophage grand écart mais sans qu'aucune d'elle ne s'avère être meilleure.

Notre étude, qui a eu lieu au service de chirurgie générale infantile du Centre Hospitalier Universitaire Mohamed 6 de Marrakech, a inclus 21 patients sur une période de 7 ans allant de Janvier 2013 à Décembre 2020.

Le poids de naissance moyen était de 2790g et le taux de prématurité était de 19%. Le taux des malformations associées était de 38%; les malformations cardiovasculaires étaient les plus fréquentes (19%). 13 patients avaient une atrésie de l'œsophage type A selon classification de Gross (62%) et 8 patients type C (38%). Tous les patients ont subi une gastrostomie initiale, 8 ont subi après une réparation définitive de l'anomalie; 3 ont subi une anastomose primaire différée, 2 une interposition colique, 2 une transposition gastrique et 1 un tube gastrique inversé. Les complications postopératoires ont été rapportées chez 67% des patients et le taux de mortalité était de 43%.

A la lumière de notre étude, nous avons démontré que le taux de mortalité est encore péjoratif dans notre contexte comparé aux pays développés. Ceci ne peut que nous inciter à fournir des efforts supplémentaires et à corriger les lacunes retrouvées dans notre prise en charge afin d'offrir une meilleure qualité de vie à nos patients.

ملخص

يمثل رتق المريء ذو الفجوة الكبيرة 5 إلى 10 بالمائة من جميع حالات رتق المريء ويشكل حالة طبية وجراحية طارئة والتي يجب تشخيصها على أبعد تقدير في قاعة الولادة . علاج هذه الحالة المرضية يتطلب تدخل أطباء العناية المركزة للأطفال وأخصائي الجراحة العامة للأطفال في مرحلة أولى، ثم على المدى البعيد أخصائي أمراض الجهاز الهضمي، أمراض الجهاز التنفسي، أمراض الأنف والحنجرة والتغذية . العديد من التقنيات الجراحية تم وصفها لعلاج رتق المريء ذو الفجوة الكبيرة إلا أن ولا واحدة منها أثبتت نجاعة أكبر .

دراستنا، التي تم إجراؤها في مصلحة الجراحة العامة للأطفال التابعة للمستشفى الجامعي محمد السادس بمراكش، تضمنت 21 مريضا على فترة 7 سنوات من يناير 2013 إلى ديسمبر 2020.

متوسط الوزن عند الولادة بلغ 2790 غرام ومعدل المرضى الخدج بلغ 19 في المائة. معدل التشوهات المرافقة بلغ 38 في المائة، تشوهات القلب والشرايين هي الأكثر شيوعا بنسبة 19 في المائة. 13 مريضا تم تشخيصهم برتق المريء بدون نسور (62 بالمائة) في حين 8 مرضى برتق المريء مرفوق بنسور (38 بالمئة). جميع المرضى خضعوا لفغر معدة أولي، بعد ذلك 8 مرضى خضعوا لإصلاح نهائي لرتق المريء ذو الفجوة الكبيرة: 3 مرضى خضعوا لمفاغرة أولية متأخرة، 2 لتوسط عن طريق القولون، 2 لتحويل المعدة و 1 لأنبوب معدي. تم رصد مضاعفات في 67 بالمائة من المرضى ومعدل الوفيات بلغ 43 بالمائة.

على ضوء هذه الدراسة، يتبين أن معدل الوفيات في سياقنا لا زال مرتفعا مقارنة مع الدول المتقدمة . وهذا لن يدفعنا إلا إلى بذل المزيد من الجهود وتصحيح مكامن النقص في طرقنا العلاجية من أجل توفير نوعية حياة أفضل لمرضانا.

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أقْسِم بِالله العَظِيم

أن أراقبَ الله في مِهْنَتِي.

وأن أصُونَ حياة الإنسان في كآفّةِ أطوَارهَا في كل الظروف والأحَوال باذلا وسْعِي في استنقاذها مِن الهَلاكِ والمرَضِ والألَم والقَلَق.

وأن أحفَظَ لِلنَاسِ كرَامَتهُم، وأسْتر عَوْرَتهُم، وأكتمَ سِرَّهُمْ.

وأن أكونَ عَلى الدوَام من وسائِل رحمة الله، باذلا رِعَايَتي الطبية للقريب والبعيد، للصالح والطالح، والصديق والعدو.

وأن أثابر على طلب العلم، أسَخِّرَه لِنَفْعِ الإِنْسَان ..لا لأذَاه.

وأن أُوَقّرَ مَن عَلَّمَني، وأُعَلّمَ مَن يَصْغرَني، وأكون أخاً لِكُلِّ زَميلٍ في المِهنَةِ الطِّبِّيَة مُتعَاونِينَ عَلى البرِّ والتقوى.

وأن تكون حياتي مِصْدَاق إيمَاني في سِرّي وَعَلانيَتي، نَقِيَّة مِمّا يُشينهَا تجَاهَ الله وَرَسُولِهِ وَالمؤمِنين.

والله على ما أقول شهيدا

كليـــــــة الـطـب
و الصيدلة - مراكش
FACULTÉ DE MÉDECINE
ET DE PHARMACIE - MARRAKECH

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جامعة القاضي عياض
UNIVERSITÉ CADI AYYAD

سنة 2021 أطروحة رقم 174

تدبير رتق المريء ذو الفجوة الكبيرة

الأطروحة

قدمت ونوقشت علانية يوم **2021/10/13**

من طرف

السيد إلياس المغاري

المزداد في 17 غشت 1995 بمراكش

لنيل شهادة الدكتوراه في الطب

الكلمات الأساسية:

رتق المريء - فجوة كبيرة - تدبير.

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