Congenital Esophageal Atresia with Tracheo – Esophageal Fistula

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التشويهات الخلقية للمرئ، ضمور المرئ مع ناسور
ما بين المرئ والقصبة الهوائية

الخلاصة

دراسة (25) حالة حديثي الولادة مصابين بتشوه خلقي ضمور المرئ مع ناسور ما بين المرئ والقصبة الهوائية في مستشفى الكاظمية التعليمي/بغداد ما بين عام 2007/2004، كما شملت (25) مريضاً, (10) منهم ذكور و(15) من الاناث وكانت نسبة الذكور الى الاناث 1.5/1 أو 3/2، وحسب تقسيم الوترستون (waterstone) كانت (18) حالة من نوع (A)، وكانت نسبة الوفاة 16%.

الهدف من هذه الدراسة هو الوقوف على الاسس في تشخيص الحالات وطرق العلاج المتبعة لقسم جراحة الصدر والاوعية الدموية وكذلك لمعرفة الاسباب في ارتفاع نسبة الوفيات .

Abstract

The records of patients with congenital esophageal atresia with tracheo–esophageal fistula at al kadhymia teaching hospital between 2004–2007 were reviewed and studied.

This comprised 25 patients (10) were male and (15) were female (M/F= 2/3).

According to waterston classification, the commonest type was type (A) 18 cases.

The basic principles in the assessment and the way of management which are adopted in our department are outlined; all were subjected to correction and primary anastomosis with hospital mortality rate of 16%.

This study is conducted in an effort to analyze the factors responsible for this high mortality rate and to discuss problems pertaining to management.

Introduction

The first account of esophageal atresia was by William Durston in 1670[1,2], and that of the esophageal atresia with tracheo-esophageal fistula was reported by Thomas Cibson in 1697[3]. Tracheo – esophageal fistula without esophageal atresia was first recognized by Lamb in 1873[4].

Dr.Thomas H.Lanman in 1940 performed the first primary extra-pleural esophageal anastomosis in a newborn with atresia of esophagus.

The classification of the varieties of esophageal atresia and associated fistula by Vogt in1923 [5], with some modification reported by Waterston et al in 1962 [5], Holder et al in 1962 [5], many of the researches have attempted to classify these anomalies and to use letter designations, which may well be more confusing than helpful. In this study we adopted Waterston classification, figure(1).

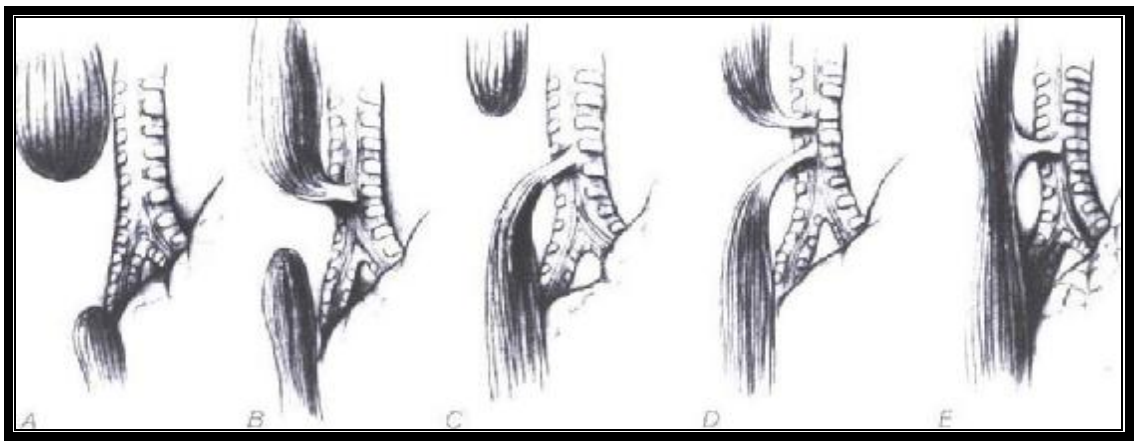

Figure 1: Waterston Classification

The etiology is unknown, and we don't really know what causes these problem and we don't think is inherited, but has familial tendency [6]. Babies with TEF and EA, have significant associated anomalies, ranging between 30-70%[5,7,8] prematurity occurs commonly, congenital TEF arise due to failed fusion of the tracheoesophageal ridges during the third week of embryological development.It often found in association with Coloboma of Iris, various heart abnormalities,choanal atresia,retardation,genital defects,and ear abnormalities an association known as CHARGE. (TEF is also a part of the VACTERL association). Cardiovascular, Anorectal anomalies, Genitourinary, Gastrointestinal, Vertebral/Skeletal, Respiratory, Genetic and Miscellaneous.

The incidence 1/3000 – 4500 live births [7].UK, USA.

The Symptoms depend basically on esophageal obstruction and secondary on respiratory tract involvement. At birth the infant has difficulty swallowing, they may spit up a lot or have lots of bubbly mucous in their mouth, eating produce severe coughing spells that interfere with breathing, aspiration pneumonia can develop from fluid breathed into the lung, small H-type fistula may go undiagnosed until later in life. The symptoms of an H_type fistula include frequent pulmonary infections and bouts of abdominal bloating.The

diagnosis is to think of the condition, passing through a nostril a semi – rigid tube, of size 8-10 French gauge, preferably incorporating a radio-opaque marker in it's wall, figure (2).The instillation of liquid contrast material few milliliter had advantages and disadvantages [9], which allowes pictures to be taken of the esophagus x-rays may show air in the bowels. Endoscopy often failes to located the fistula if it small [10]. Radiography was traditionally used,three-dimensional CT scan coupled with reformation in the three orthogonal planes may have a complementary diagnostic role in congenital esophageal atresia [11,12].

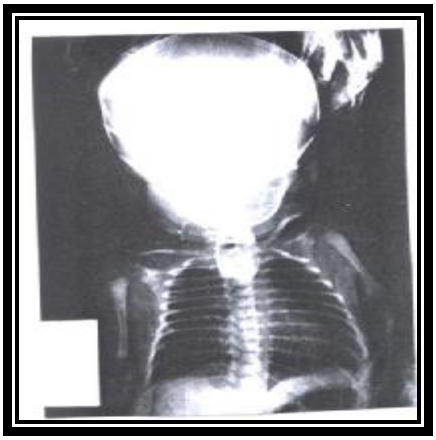

Figure 2: Type A EA/TEF. Catheter in upper pouch. Gas below diaphragm.

Classification of infants on the basis of factors likely to influence survival into three groups [13].

1 - Group A: Over 5.5 Ib (2.27 Kg) birth weight and well.

2 - Group B: Birth weight 4-5 Ib (1.8 – 2.27 Kg) and well.

Higher birth weight, moderate pneumonia and moderate congenital anomaly.

3 - Group C: Birth weights under 4 Ib (1.8Kg).

Higher birth weight, severe pneumonia and severe congenital anomaly.

Treatment, surgery, but surgery cannot always be performed immediately because of prematurely, the presence of other birth defects, or complications from aspiration pneumonia. It is usually done it a hospital that has special facilities for treating seriously ill newborns.

The operative management either one stage or staged operation, depend on the long gap, prematurity, associated anomalies [14,15].

While awaiting surgery, the infant's condition is stabilized. Preoperative care concentrates on avoiding aspiration pneumonia and includes:

* Elevating the head to avoid reflux and aspiration of the stomach contents.
* Using a suction catheter to continuously removed mucus and saliva that could be inhaled
* When necessary, placement of a gastrostomy tube.
* With holding feeding by mouth.

Thoracocsopic repair of EA and TEF represents a natural evolution in the operative correction ff this complicated anomaly and can be safely performed by experienced endoscopic surgeons, especially benefit for babies based on the associated musculoskeletal problems following thoracotomy [16,17].

After surgery,the infant will be cared for in a hospitals neonatal intensive care unit (NICU) the infant will be placed in a machine called an isolette (incubater) to provide warmth and prevent infection. The treatment includes:

* Oxygen; Breathing assistance; Mechanical ventilation; Intravenous fluid; Antibiotics as needed; Pain medicine as needed.

A tube nasogastric keep the stomach empty. Feedings are started through this tube as soon as bowel function starts again.

The TEF are not preventable birth defects.

Materials and Methods

This is a study of (25) patients with EA with or without TEF, from august 2004 to march 2007.

All cases were managed at the department of thoracic and cardiovascular surgery unit of Al kadhymia teaching hospital in Baghdad.

Fifteen patients (60%) were girls and ten (40 %) were boys.

Twenty one (84 %) of admitted patients were from urban region and four (16 %) from rural region.

Twenty-four patients delivered at hospital, (7) normal vaginal delivery, (17) with caesarean section .The other (1) patient delivered at home.

A similar anomaly could not be traced in the family of our patients.

Results

The trachoesophagel fistula and esophagel atresia patients were admitted directly to our unit or referral from padiatric hospital, central child hospital, or from outside Baghdad. Their birth weight ranged from 1700-4350 grams, table(1).

Birth weight / grams	No. of patient	Percentage %
< 1800 g	3	12 %
1800 – 2500 g	9	36 %
> 2500 g	13	52 %
Total	25	100%

Table 1: Birth Weight

And prematurity was seen in 20 % of cases, table (2).

Maturity of baby	No. of patients	Percentage
Premature	5	20%
Mature	19	76 %
Post-mature	1	4 %

Table 2: Body Size

The type of esophageal atresia with tracheo-esophageal fistula are shown in the table (3).

Type	No. of patients	Percentage
A	18	72 %
B	5	20 %
C	///	///
D	1	4 %
E	1	4 %

Table 3: Types of EA/TEF based on Waterston classification.

The age incidence of these patients at the time of admission more during 1- 4 days of age, about (14) (56%), table (4).

Age of patients Age/day	< 1day	> 1-4day	5-10d	11-30d	31-1y
No. of patients	4	14	5	1	1
Percentage	16%	56%	20%	4%	4%

Table 4: Age incidence of admission

Other associated congenital anomalies founded in (9) patients, Vertebral defect (0), Anal anomalies (3), Cardiac anomalies (4), Tracheopulmonary (1), Radial limb (0), Facial defect (1), CNS & Down's Syndrome, Renal defect, Bowel atresia, abdominal wall defect cannot be detected in these cases.

Seven patients (28 %) associated with moderate pneumonitis mainly of right upper lobe and two patients (8 %) with severe pneumonia, detected clinically and radiologically.

Three patients had pre-natal complication, was diagnosed immediately after birth, but unfortunately is only (1) case immediate surgical intervention was performed.table (5).

Age of patients Age/day	< 1day	> 1-4day	5-10d	11-30d	31-1y
No. of patients	3	15	5	1	1
Percentage	12%	60%	20%	4%	4%

Table 5: Age at time of operation

Although these cases are considered as emergency yet in only (2) patients operation done on the same day of admission. Table (6).

Time/day	Same/day	1-2	3-4	5-6	7-8	9-12	Month	Year
No. of patients	2	3	17	1	1	0	1	0
Percentage	8%	12%	68%	4%	4%	0%	4%	0%

Table 6: Time of operation

The various surgical techniques is based on joining (sewing) together the two ends of the esophagus (an anastomosis) using fine suturing. If a TEF is present this must be repaired first and the hole with trachea closed. About (80%) the operation is straight forward and completed satisfactorily.

The long gap situation, however, the operation may be much more difficult and results not always good. The two ends of esophagus may be thought to be too far apart, or the tissue too thin, raising concern that the repair would be under too much tension and not hold up.

For gaps 2-3 cm long or 2-4 vertebral bodies apart, a primary repair will usually be carried out. Surgeons are realizing that some tension will be tolerated by a well constructed anastomosis. As the distance increases, however, so do the likelihood of complications with an attempted primary repair. To avoid this possibility by doing staged operation, while we wait:

A - Stimulating the esophagus to grow 12-14 days [18].

B - Upper pouch has been brought out the neck (a spit fistula).

C - The traction sutures are placed in the esophageal ends and brought through the skin to the outside of the chest wall. This allows the traction to be increased daily and maximizes the growth stimulus. These children kept on the ventilator and heavily sedated so they do not tear the traction sutures loose.

D - Esophageal substitutions (interposition grafts), gastric transposition or stomach tube. Colonic interposition; but suffice it to say, the likelihood of a difficult early course is high, (20%) staged operation, (12%) esophagestomy and feeding gastrostomy, (4%) done stomach tube, (4%) colonic intreposition.

One patient had H-type fistula, repaired done through right coller incision, no other type of surgery done.

The commonest post-operative complications are respiratory in origin caused by pneumonia or heart failure, table (7).

Type of Complications	No. of Patient	Percentage
Sepsis (infection)	1	4 %
Renal failure	0	0%
Tracheomalacia	1	4%
Respiratory distress	4	16%
Dehydration	2	8%
Leakage	0	0%
*Minor	0	0%
*Major	0	0%
Esophageal stricture	2	8%
Esophageal dysmotility	1	4%
Heart failure	2	8%
Recurrent fistula	0	0%
Food impactation	0	0%

Table 7: Postoperative complications

One of most serious immediate complication is leakage at the site of anastomosis, and most common remote complication is stricture and recurrent fistula.

Minor leak by conservative management:

A - Intravenous fluid.

B - Nasogastric tube inserted under screen guide.

C - Cover by broad spectrum antibiotics.

D - Observation.

While major leak treated by resuturing and feeding gastrostomy tube. Stricture treated by dilatation through esophagoscope either single time or multiple at 2-month, 6-month and1-year.

The mortality rate was 16%, table (8).

Sex	No. of living patients		No. of death patients	
Female	13	25%	2	8%
Male	8	32%	2	8%
Total	**21**	**84%**	**4**	**16%**

Table 8: Mortality rate

The neonatal deaths by Waterston risk group, table (9).

Group	No. of patients		Mortality rate		Survival rate	
Category A	18	64%	0	0%	18	64%
Category B	3	12%	2	8%	1	4%
A	2	8%	1	4%	1	4%
B	1	4%	1	4%	0	0%
Category c	4	16%	2	8%	2	8%
A	2	8%	0	0%	2	8%
B	2	8%	2	8%	0	0%
Total no.	**25**	**100%**	**4**	**16%**	**21**	**84%**

Table 9: Mortality rate based on Waterston risk group

Discussion

This study done on patients treated at Al-kadhymia teaching hospital between 2004-2007.

These records were reviewed and studied. to present a fairly representive picture of this anomaly in Iraq as this center is one of the main referral centers of such cases from various parts of the country to examine various methods of diagnosis that are presently performed, to discuss the reasons for high mortality rate, and to analyze the problems encountered in the management.

The baby's condition before these large operation is important. But in addition to the baby's general condition, there are at least three important issues which will affect the treatment plan. These are whether or not the baby:

1 - Is very premature.

2 - Or has other serious birth defects in the heart, kidney, or elsewhere or,

3 - Has a type of esophageal atresia that will be difficult to repair.

These are potentially complicating issues that may significantly affect the proposed operation in terms of either timing or final result.

While the surgery of an infant or a small child requires certain consideration which differs from those of surgery in adults, it is obvious that surgery in a premature baby calls for special care and delicacy because of smallness and fragility of the tiny subject and also because of physiological variations which are known to exist.

While the combination of prematurity and a serious anatomical defect might seem to be overwhelming, and even incompatible with survival. Unfortunately ideal hospital facilities and prompt care are not available in our center.

This ratio of M/F is 2/3, and no a similar anomaly in the family could be detected. It was found is commoner among multipara, and in previous history only one mother was subjected to radiation. The presenting symptoms are classical in all patients and include coughing, choking and cyanosis on feeding. The associated aspiration pneumonitis due to excess salivation and regurgitation are the main feature in delayed cases.

Patients whose diagnosis was made during the first day of life have very poor survival rate than those diagnosed during the second or third day of life. This is because these babies have sever congenital anomalies associated with EA/TEF and they are so sick when presented in early hours of life after birth, so that they carry high morbidity and mortality rate than other who diagnosis early without any associated anomalies who had good prognosis.

This statement should in no way be misunderstood that an early diagnosis is not only important. But Operations should be performed as soon as possible but not until dehydration is corrected and pneumonitis is treated. Baby who comes to operation poorly prepared may not only suffer complications but also carry a high rate of mortality.

Whatever operative approach is practiced, the shorter the duration of time the premature infant is subjected to anesthesia and operative trauma, the better are his chance for survival. Some associated congenital anomalies may require surgical relief prior to or at the time of surgical correction of esophageal obstruction.

Advance which have been made in operative and the postoperative care of infants have contributed greatly to the increasing success in this form of surgery. No circular cut through the muscle wall will allow the remaining tissue to stretch and balloon up to a serious degree and always kept stomach below diaphragm. Overall goals have successful true primary repairs, those who are far enough out from repair (usually a year), to allow complete resolution of anastomotic strictures and to have overcome oral aversion, and eating normally. 30% may have gastro esophageal reflux after repair and need fundoplication for significant (GER). For the very long term, we do not fore see any problems with the esophagus after a true primary repair, the function of their esophagus appear to be very satisfactory. All patients are kept under close watch in pediatric intensive care unit. Postoperative complications in infants can occur so rapidly

in such that, extra-precautions must be taken and so they should receive constant extra-nursing care.

Risks for any anesthesia:

A - Reactions to medications.

B - Problems of breathing.

Risks of surgery:

A - Bleeding.

B - Infection.

Additional risks include:

A - Narrowing of the repaired organ.

B - Collapsing lung (pneumothorax).

C - Hypothermia.

Such intensive care and nursing care unit are deficit or even not available in our hospital.

A comparative study of mortality rate between our center and other shown in table (10).

Study	Group A		Group B		Group C	
Our study	**Survival**	**Death**	**Survival**	**Death**	**Survival**	**Death**
	18	0	1	2	2	2
	18		3		4	
Total	25					
Holder study (11)	37	1	23	2	25	12
	38		25		37	
Total	100					
Grosfield (12)	13	0	40	3	13	4
	13		43		17	
Total	73					

Table 10: Waterston risk group (Comparative Study)

The results are disappointing and this may be due to respiratory problems and associated anomalies, and also due to:

1 - Late diagnosis.

2 - Late referral.

3 - Lacking ideal diagnostic facilities to detect congenital anomalies.

4 - Improper preoperative assessment and preparation.

5 - Lacking paediatric intensive care unit.

6 - Shortage or inexperienced anaesthetist.

7 - Massive causality makes less care to cold cases.

8 - Collapsing in the system of medical and paramedical staff.

Conclusions

We have seen that the true primary repair is possible, apparently in all (TEF/EA) patients.

This esophageal repair will reliably allow the children to eat normally and not be dependent on a gastrostomy tube.

We believe the benefits of this approach will only increase with time. Seems to be no evidence for late problems arising with esophagus however, the potential for difficulties from untreated (GE) reflux remain. Cleary, however, after few years, even with the very long gap repairs cannot be distinguished from the normal esophagus by what they eat.

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