EPIDEMIOLOGY OF ESOPHAGEAL ATRESIA: A HOSPITAL- BASED STUDY IN SULAIMANI

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ABSTRACT

Background

Esophageal Atresia (AE) is a sporadic unpreventable birth defect, consisting of lack of continuity between the upper and lower esophageal pouches, with prevalence rate 1-5/10 000.

Objective

The aim of this study is to describe some epidemiologic data on esophageal atresia, estimating it's prevalence in Sulaimani province, and characterizing the cases with regards to age at presentation, gender, gestational age, pathological classification, associated anomalies, postoperative complication, and mortality rate.

Methods

This study extended from June 2006 to December 2014, in which 100 patients with esophageal atresia were managed in Pediatric Surgical Unit of Sulaimani Teaching Hospital in Sulaimani province.

Results

the total number of cases was 100 patients (61 males and 39 females), the mean gestational age was (37) weeks, and 56% had birth weight > 2500 gm. The most common type was [EA] with [TEF] (92%), thirty two percent presented between 3-7 days of age. Sixty percent of the patients developed postoperative complications; the overall birth prevalence rate in Sulaimani was 4.7 cases per10 000 live birth. The mortality rate was (69%).

Conclusion

Esophageal Atresia is a rare disease in Sulaimani, with the most common type being Esophageal Atresia with distal tracheoesophageal fistula. Unlike other studies, in the age of presentation, those presented after the first week, had higher survival rate.

Keywords: *Esophageal Atresia, Tracheoesophageal fistula, Epidemiology, Sulaimani.*

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INTRODUCTION

Esophageal Atresia (EA) is a sporadic unpreventable birth defect [1]. The recorded history of EA and Tracheoesophageal fistula (TEF) dates back to the 17th century. With incidence estimated to be 1 case in every 2500-3000 live births and associated with TEF in 90% of the cases [1, 2]. Both situations are the result of a failure in the division of the anterior primitive gut, in the formation of the trachea up front and the esophagus behind forming two separate tubes that normally take place between 3rd and 5th week of development, and is completed in week 8th [1, 3].

Because most EA malformations occur sporadically, it is highly unlikely, that this is a simply inheritable condition. The pathogenesis is therefore most likely heterogeneous and multifactorial and involves multiple genes and complex gene-environment interactions [1,4]. Various environmental factors have been suggested as risk factors, studies from the European Surveillance of congenital anomalies birth registry network found that older mothers are at a significantly greater risk of having a child with EA [5]. Several classifications have been advocated but all are physiological [6]. The Spitz classification is suitable to use for preoperative predictor to parental counselling and comparing of treatment outcomes of EA among pediatric tertiary care centers [7].

Up to 50-70% of the children affected by this condition also present with associated congenital abnormalities or chromosomal anomalies 12, 13 [8].

The effect of EA on the infant is aspiration of saliva or milk, soiling of the lung. Pulmonary complication may follow, initially atelectasis and then pneumonitis [1, 4].

Sometimes EA is diagnosed on antenatal ultrasonography [6, 9], a morphologic and karyotype usually done after reaching a prenatal diagnosis. Nowadays ,the diagnosis in most cases established immediately after birth [1, 10]. It's suspected when a new born infant appears to be drooling excessively, and the diagnosis is confirmed when an orogastric catheter cannot be passed through the mouth and esophagus into the stomach and typically stops at 10-12 cm [1, 11, 12].

X-Ray of the thorax and abdomen is important to confirm the position of the tube, provide information about the cardiac silhouette and pulmonary infiltrate, the location of the aortic arch and the presence of vertebral and rib anomalies. Air in the stomach confirms the presence of a distal fistula [12]. Echocardiography and renal ultrasonography should be done to exclude associated abnormalities [1, 12, 13].

The operative procedures differ according to the pathological type. In 1941, Haight performed the first successful primary repair for EA with distal TEF [1]. The prognosis of EA with or without TEF will depend on presence of associated malformations [1, 12, 13], it is also influenced by birth weight and development of aspiration pneumonia due to delayed diagnosis . Prompt diagnosis, appropriate clinical management and expeditious referral to a tertiary care center have a dramatic impact on the improved survival of these infants. Estimates today suggest that, in the absence of other severe anomalies, survival rates in these infants approach 100 % [14, 15]. The aim of this study, is to describe the important epidemiological data on EA in Sulaimani including age at presentation, the sex incidence ,the mean gestational age, the mean birth weight, to find out the common type of EA, the abnormality commonly associated with EA, to find out the mortality rate and the factors impact on this rate, finally to measure the live birth prevalence .

PATIENTS AND METHODS

This study analyzes 100 neonates, admitted to the Pediatric Surgical Unit from June 2006 to the end of December 2014, with three variants type of esophageal atresia and /or tracheoesophageal fistula. Data were collected prospectively and included age and sex, birth weight, pathological type, methods of investigation, definitive treatment and outcome. Data was entered into excel data sheet and statistical analysis was performed, using Chi square test, P value <0.05 was regarded as significant.

History of maternal polyhydromnia was positive in most of cases but because it have low positive predictive value we didn't depend on it for antenatal diagnosis .In this study EA was detected in two patient prenatally, the first was EA with TEF detected by ultrasound,

showing the upper neck pouch sign associated with polyhydromnia, the second was a case of pure EA and diagnosed by ultrasound showing gasless abdomen associated with polyhydromnia.

Postnatal diagnosis mostly depended on the typical clinical presentation such as, excessive salivation, cough, apnea, tachypnea, and cyanosis when feeding was attempted, also the patient's inability to pass a

rigid radio opaque nasogastric tube from the mouth to the stomach was diagnostic for EA and/or TEF, but these finding were confirmed with radio-graphic visualization of the tube coiled in the proximal pouch (Figure 1). Preoperative investigations included; Plain Chest X-R radio graphs which provide much information rather than confirmation of EA ,including findings for and depiction of the position of the aortic arch, as well as the presence of any vertebral or other associated anomalies (Figure 2), The length of the esophageal gap is usually not known preoperatively. In EA/TEF, a longer gap between the two esophageal ends should be expected when the distal fistula is found at the carina combined with a short upper pouch, or in case of pure EA with gasless abdomen (Figure 3), where long gap exists between the two esophageal segments and may not be amenable to an initial primary repair.

Echocardiography should be performed prior to operation, as it may reveal cardiac and/or aortic arch anomalies. A right descending aorta, which occurs in about 2.5% of the cases, may make a left-sided thoracic approach preferable.

Generally, the operative treatment of EA/TEF is not regarded as an emergency procedure. Thus, there is usually time to confirm the diagnosis and to assess for associated anomalies. Measures were being taken

to reduce the risk of aspiration. The oral pharynx should be cleared, and frequent done, the infant's head kept elevated, intravenous fluids started, and Oxygen therapy was used as needed to maintain normal oxygen saturation. In infants with respiratory failure, endotracheal intubation performed. If sepsis or pulmonary infection was suspected, broad-spectrum antibiotics (such as ampicillin plus gentamicin) administered. Some surgeon, however recommend starting intravenous antibiotics empirically because of the increased risk of aspiration.

All of the children were operated by a pediatric surgeon, as a special type of operations. The cases of EA that were associated with TEF were opened through right posterolateral thoracotomy (4th intercostal space) incision in the left lateral decubitus position, via an extra pleural approach, identification of the upper and lower pouch with the fistula (were divided and the tracheal end were over sewn) .A single layer anastomosis was performed in all of them, leaving a chest tube behind for one day more after starting oral feeding. Contrast-swallow (water soluble) was done at the 6th postoperative day for checking the patency (Figure 4).

The babies with pure EA had a long gap esophageal atresia and represents a challenge, delayed repair were done with feeding gastrostomy and cervical esophagostomy followed by colonic interposition.

RESULTS

Among the 100 patients diagnosed as EA during the last 9 years, 61% of the patients were male and 38% were female patients. The mean birth weight was 2125 gm (56%) of the patients had birth weight more than 2500 gm, while only 12% were less than 1800.as shown in figure 5. The mean gestational age was 37weeks (range, 29 to 39 weeks), as shown in figure 6, that 88% were full term. The most common type was EA with distal TEF in 92cases (92 %), seven cases with pure EA (7%), one case of EA with proximal and distal fistula (1%) as shown in Table 1. Regarding the age at presentation, we had received these cases usually after the first week of life, but in the last few years because of increased knowledge among the junior pediatrician working in the Sulaimani districts and sub-districts, they were diagnosed more promptly, even few hours after birth, with highest proportion between 3-7 days of life, as shown in Table 2.

The presenting symptoms are classical in all those patients like breathing problems, frothy secretion from mouth, coughing, choking and even cyanosis. Associated congenital anomalies were found out in 65 % of cases, the most frequent was congenital heart disease, among which patent ductus arteriosus was the most common anomaly, followed by imperforate anus, VACTERL syndrome and small bowel atresia, shown in Table 3.

Postoperative complication developed in 60%. Anastomotic complication including; minor leakage occur in 12 patient which treated conservatively, and resulted in death of 3 patients. Major leakage in two patients treated conservatively in one, and need revision in the second (Figure 7), with very good outcome in both.

The higher survival rates were recorded in those patients presented at the age of one week or more, similarly the lowest mortality rate was in that age group. This result was statistically significant (p=0.02), as shown in (figure 8) and table 5.

The overall mortality rate was 69 %, 62 % post operatively, 5% during the operation and 2% preoperatively, as are shown in Table 6.

The live birth prevalence rate was 4.7 per 10 000; the mean incidence of EA during the last 8years was (0.0005).

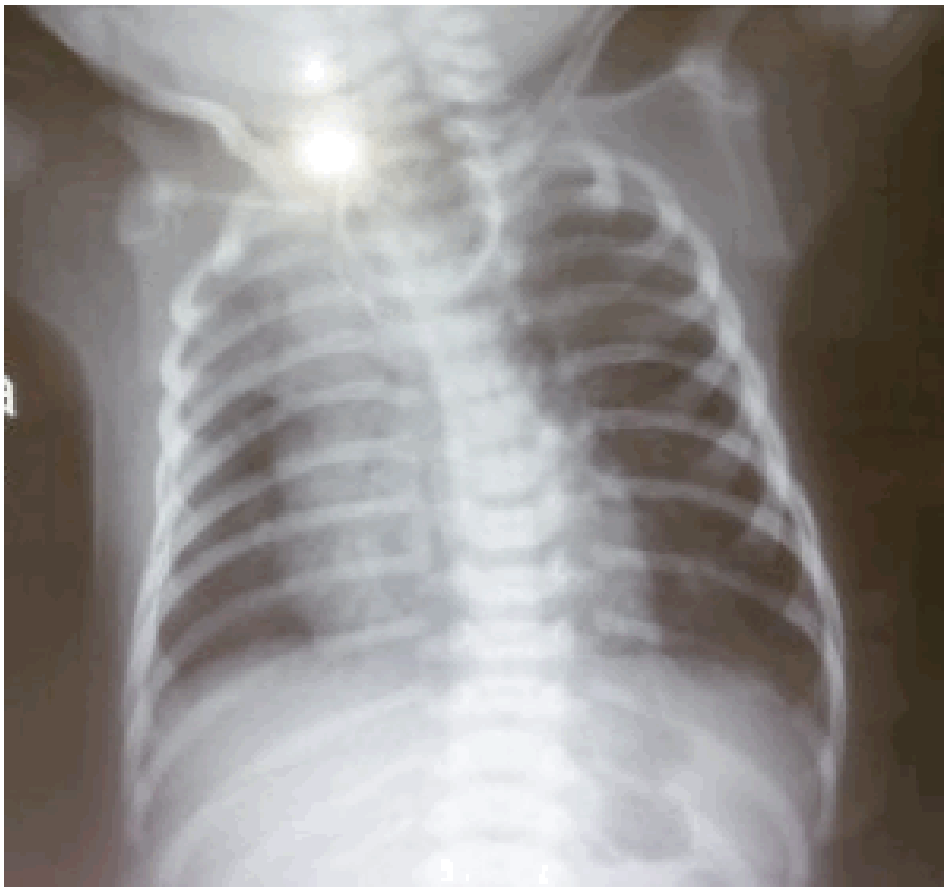

Figure 1. Radiographic visualization of the nasogastric tube coiled in the proximal pouch.

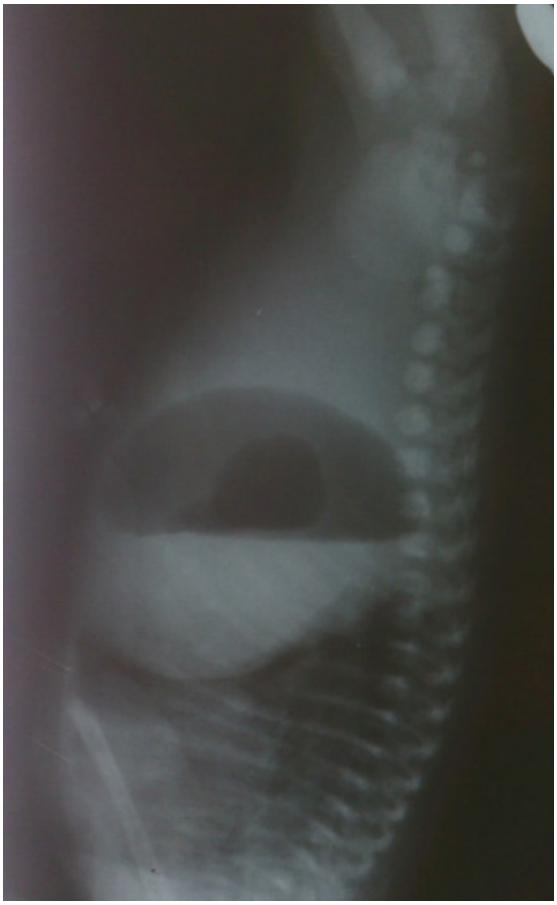

Figure 2. A case of Tracheoesophageal Atresia associated with Imperforate anus and small bowel atresia.

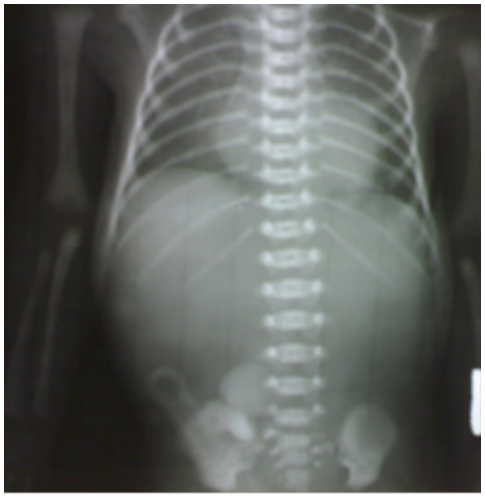

Figure 3. Gasless abdomen in Pure Esophageal Artesia.

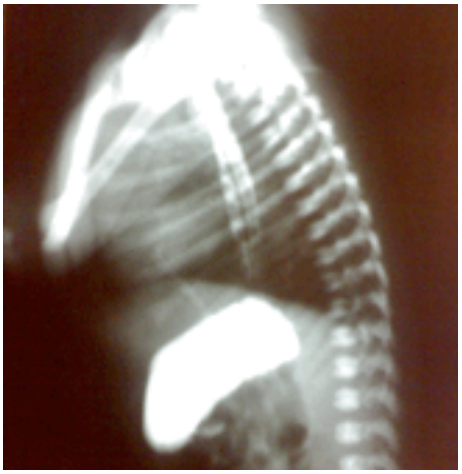

Figure 4. Contrast swallow to check the anastomosis post operativelypatent and intact).

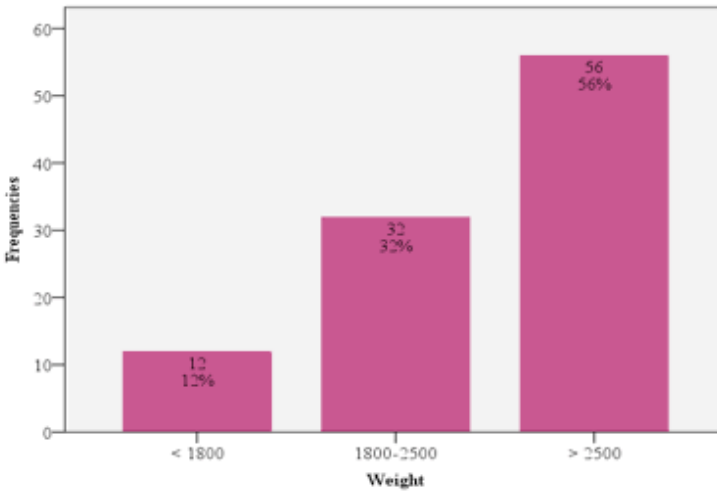

Figure 5. The Birth Weight.

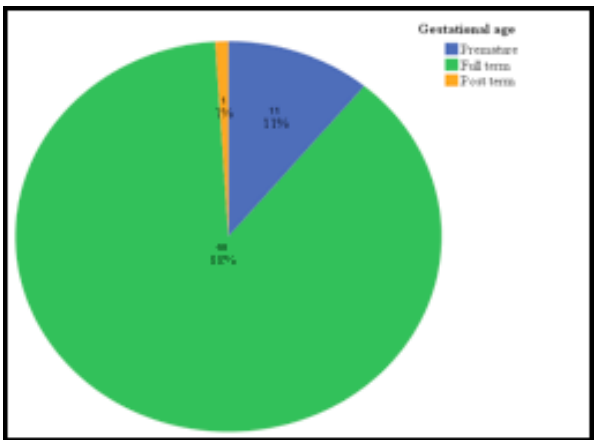

Figure 6. The Gestational Age.

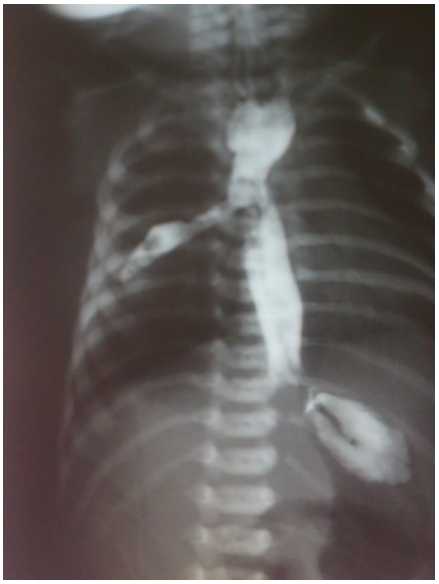

Figure 7. Contrast swallow, shows major leak.

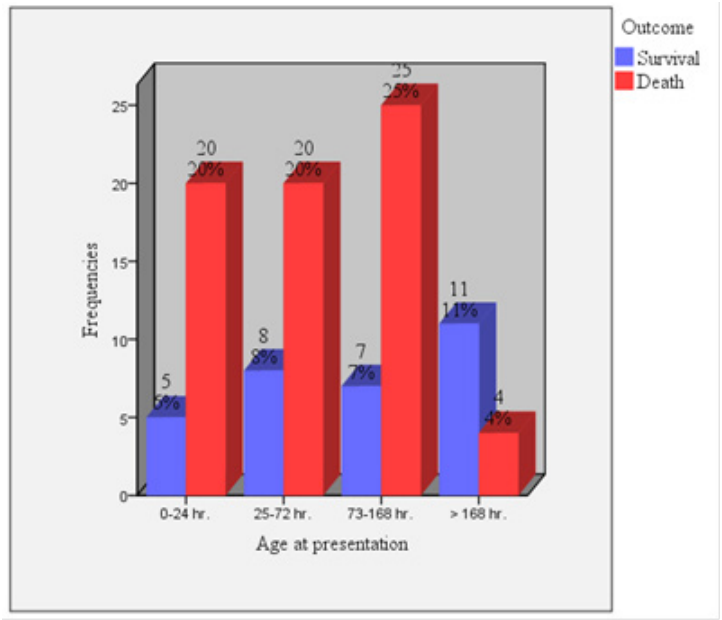

Figure 8. Relation between the outcome and the age of presentation.

Table 1. The Pathological Types of EA.

Type of EA	Frequency	Percent
EA with distal TEF	92	92
Pure EA	7	7
EA with prox. & distal TEF	1	1
Total	100	100

Table 2. The Age at the time of presentation.

Age at presentation	Frequency	Percent
0-24 hr.	25	25%
25-72 hr.	28	28%
73-168 hr.	32	32%
> 168 hr.	15	15%
Total	100	100%

Table 3. The Associated Anomalies.

Associated anomalies	Frequency	Percent
CHD	62	62%
IA	6	6%
Down syndrome	2	2%
VACTERL	2	2%
Bowel atresia	1	1%
Total	73	73%

Notes:

-Some patients had more than one associated anomalies.

-Congenital heart disease (CHD), Imperforate Anus (IA), Vertebral, Ano-rectal, Cardiac, Tracheal, Esophageal, Renal and Limb anomalies (VACTERL).

Table 4. Post operative complications.

Post-operative complication	Frequency	Percent
Minor leak	12	12%
Major leak	3	3%
Stricture	8	8%
Sepsis	20	20%
Resp. failure	22	22%
Cardiac failure	1	1%
Food impaction	2	2%
Renal failure	2	2%
Total	66	64%

Table 5. Relation between the outcome and the age of presentation.

Age at presentation	Early outcome		Total
	Survival	Death	
0-24 hr.	5	20	25
25-72 hr.	8	20	28
73-168 hr.	7	25	32
> 168 hr.	11	4	15
Total	31	69	100

Table 6. Time of death.

Time of death	Frequency	Percent
Preoperative	5	5.0
During operation	2	2.0
Postoperative	62	62.0
Total	69	69.0

DISCUSSION

This study was done on 100 patients with EA at pediatric surgical department in Sulaimani, to present the important epidemiological facts of this congenital malformation, and the outcome of our management, as this center is the only referral center for such anomaly in this province. The limitation of the study was inability to investigate for the geographical differences, which may provide an insight into the underlying etiology. It appears to be a slight male predominance, although this also appears in this research but this is not a universal finding and may not be true for all the varieties [1, 16].

The mean birth weight was 2125 g, which is similar to other studies [16]. The mean gestational age was 37 weeks (range, 29 to 39 weeks) also similar to other study [17].

Aspiration pneumonitis due to excess salivation and regurgitation were the main feature in delayed referred cases [1, 18]. Most of these cases had EA with distal TEF which was found in 92%, referring to the literatures it is also the most common type, but found to be 85-86% [1, 12, 14, 15, 16], this difference could be explained by exclusion of the one of the five main pathological type,TEF without EA, whose managed usually in Cardiothoracic center. Pure EA was the second common type, and this is also described in the classification of EA in most of the literatures [1].

Currently the associated anomalies may determine survival, and alter the treatment approach [14, 15]. In this study they were present in 65% of cases, which is similar to other studies [1, 15]. Congenital heart disease was the most common associated anomaly (62%) ,like other studies, but with lower incidence [1, 13, 14, 15], followed by imperforate anus (6%) also similar to other published research [15], followed by VACTERL syndrome (2%), with no significant difference from literature [13].

The most common age of presentation was between 3-7 days, and the most of the survived patients were those operated at age of more than one week, and those were presented and diagnosed within the first few hours after birth had lower survival rate .Although this was against most of other studies, but it was noticed also in a study done in Baghdad [19]. This result was statistically significant (p=0.02). This explained by a fact that those with poor prognosis usually diagnosed then operated earlier, while those having better prognosis presented and managed later and having higher survival rate.

The three priorities in the treatment are to save life, to achieve alimentary continuity and to preserve the esophagus. But the management is affected by established pneumonia, and associated anomalies [1, 14], in addition to prematurity and low birth weight. All the patients are kept under close observation in pediatric surgical unit. Advances which have been made in operative and the postoperative care of neonates have contributed greatly to the increasing success in this form of surgery.

Nowadays, all patients with EA are expected to survive unless there are major congenital malformations affecting other system in most of developed countries [1]. In a research conducted in USA, the overall survival rate was 95% [15]. But survival rate was low (31%) in our department. This can be explained by delay diagnosis (although this has greatly improved in the last five years), lack of pediatric intensive care unit for receiving these patients post operatively, and lack of total parenteral nutrition for those with long period on nothing by mouth.

The overall prognosis of the survived patient is good, eight of them during the data collection period required endoscopic dilatation, all of them developing recurrent

respiratory tract infection possibly due to repeated minimal aspiration occurring with gastroesophageal reflux (GER) and esophageal dysmotility which is a long term post-operative complications [20]. Most of the patients were from district or sub-district which is against another study, with higher incidence in urban area [16], but this may attributed to variability in population ethnic composition as mention in a study done in Europe [21]. A similar anomaly didn't trace in the families of our patients.

The overall birth prevalence rate in sulaimani was 4.7 cases per 10 000. Ophanet, a consortium of European partners, lists Esophageal Artesia as a "rare disease" with prevalence rate 1-5/10 000 [22]. A recent study demonstrated that the prevalence of EA varied from 1.27 to 4.55 per 10 000 according to different European regions, which is close to our rate [23]. but different from other two studies done in Baghdad in the same center for different period (they didn't measure the prevalence rate, but able to collected 100 cases in the both studies during two years), although it is one of many other pediatric surgical centers in Baghdad, which are receiving similar cases [24, 25].

From our study of 100 cases of EA with or without TEF we conclude that EA is a rare disease in Sulaimani and the most common type was EA with distal TEF; congenital heart disease was the most common associated anomalies. Mortality and morbidity is influenced by the associated anomalies, prematurity, and established pneumonia. Despite the improvement in survival now by perfect knowledge of anatomy and meticulous, skilled dissection by the pediatric surgeons (no survival cases were recorded previously in this city), the outcome in these patients still very low compared to other center. Improvement are based on early diagnosis and timely reference, good preoperative resuscitation, good anesthetic care and close follow up post operatively in an ICU.

REFERENCES

1-Rothenberg S.S, Esophageal Atresia and Tracheoesophageal Malformation In: Holcomb G.W, murphy P.J, Ostlie D.J, Aschraft's pediatric surgery, 6th edition, United Kingdom /London: Saunders/Elsevier; 2015, ch.27.365-377.

2- Spitz L, Oesophageal Atresia .Orphanet Journal of Rare Diseases 2007; http://www.OJRD.com/content/2/1/4 doi:10.1186/1750-1172-2-24.Acceessed May 11, 2007.

3- Ulusoy M, Kıvrak A.S, Uysal I.I, Karabulut A.K, Paksoy Y, Fazlıogulları Z. Developmental Anomalies of Bronchial Tree :A Multidetector Computerized Tomography Study.Int. J. Morphol. 2013; 31(3):1049-1055.

4- Brunner HG, Bokhoven V.H. Genetic players in esophageal Atresia and Tracheoesophageal Fistula. Curr Opin Genet Dev.2005; 15(3):341-347.

5- Jang EM, Felix J F, Klein AD. Etiology of Esophageal Atresia and Tracheoesophageal Fistula:" Mind the Gap". Curr Gastroenterology rep. 2010; 12(3):215-222.

6- Upadhyaya VD, Gangopadhyaya AN, Gupta DK. Prognosis of Congenital Tracheoesophageal fistula with Esophageal Atresia on the basis of gap length. Pediatr Surg Int.2007; 23(8):767-771.

7- Niramis R, Tangkhabuanbut P, Anuntkosol M, BuranakitjaroenV, Tongsin A, Mahatharadol V. Clinical Outcomes of Esophageal Atresia: Comparison Between the Waterston and the Spitz Classifications. Ann Acad Med Singapore.2013; 42(6):297-300.

8- Smith Ch Sh. Genetic factors in esophageal atresia, trachea-esophageal fistula and the VACTERL association: Roles for FOXF, and the 16q24.1Fox transcription factor gene cluster, and review of literature .Eur J Med Genet.2010; 53(1-3):6-13.

9-Kalish RB, Chasen ST, Rosenzweig L. Esophageal atresia and tracheoesophageal fistula: The impact of prenatal suspicion on neonatal outcome in a tertiary care center. J Perinat Med.2003; 31(2):111-114.

10- Kaber R, Pillai K. Esophageal Atresia/ Tracheooesophageal Fistula. In: Myers JA, Millikar KW, Saclarides. Common Surgical Disease: An Algorithmic Approach to problem solving.2nd Ed, New York: Springer; 2008.ch64.255-265.

11- Ricketts R .Esophageal Atresia and Tracheooesophageal Fistula. In: Chung D H, Chen M Y .Atlas of pediatric surgical technique. Philadelphia: Saunders; 2010.ch5.44-50.

12- Clarck DC. Esophageal Atresia and Tracheoesophageal Fistula .Am Fam Physician. 1999; 59(4):910-916.

13-Eghbalian F, Monsef A, Mousavi SH. Urinary Tract and other Associated Anomalies in Newborns with Esophageal Atresia .Urology Journal.2009; 6(2) : 9-13.

14-Spencer W, Beasley, Myers AN. Trends in mortality in oesophageal atresia .Pediatric Surgery I nternational.1992; 7(2):86-89.

15- Engum AS, Grosfeld JL, West kW, Rescorla FJ. Analysis of morbidity and mortality in 227 cases of Esophageal Atresia and/ or Tracheooesophageal Fistula over two decades. Arch Surg.1995; 130(5):502-8.

16-Pedersen R, Husby S, Garne E. 443 Epidemiological characteristics of esophageal atresia in Europe, 1222 cases. Pediatric Research.2010; 68(10):227-228.

17-Lautz TB, Mandelia A, Radhakrishnan J,VACTERAL association in children undergoing surgery for esophageal atresia and anorectal malformations. J Pediatr Surg.2015;02.049.www.ncbi.nlm.nih.gov/PubMed/25913268.Accessed Mar 10, 2015.

18-AL-Salem AH, Tayeb M, Khogair S, Roy A,AL-Jishi N,AL-Senann K. Esophageal Atresia with or without Tracheoesophageal Fistula Success and failure in 94 cases. Ann Saudi Med. 2006; 26(6):116-9.

19- Muhieldeen S. A Congenital Esophageal Atresia with Tracheo –Esophageal Fistula . AJPS .2008; 5(1):102-112.

20- Konkin D H, O'hali W A, Webber E M. Outcomes in Esophageal Atresia with Tracheoesophageal Fistula. Journal of pediatric surgery.2003; 38(12):1726-1729.

21-Nassar N, Leoncini E, Arteago VJ, Bakker MK, Bower C, Gastilla EE, et al. Prevalence of Esophageal Atresia among 18 International birth defects surveillance programs. Birth Defects Res A Clin Mol Teratol.2012; 94 (11):893-9.

22- Sfeir R, Michaud L, Salleron F, Gottrand J. Epidemiology of Esophageal Atresia. Dis Esophagus Journal .2013; 26(4):354–355.

23-Pedersen RN, Calzolari E ,Husby S ,Garne E. Oesophageal Atresia: Prevalence, Prenatal diagnosis and associated anomalies in 23 European region. Arch Dis Child.2012; 97(3):227-33.

24- AL-Kawaz Sh, Ejrish A. Esophageal atresia and Tracheoesophageal Fistula: A Study of 100 Cases. Medical Journal of Babylon.2010; 7(1-2):215-220.

25- AL-Obaidi S, Khalaf A, Abdul-Hassan A. ESOPHAGEAL ATRESIA AND TRACHIOESOPHAGEAL FISTULA. A thesis Submitted to the Iraqi commission for medical specialization in partial fulfillment of the requirement for the degree of fellowship of the Iraqi commission for medical specialization in pediatric surgery.2008.