SINGLE CENTRE EXPERIENCE OF ESOPHAGEAL ATRESIA IN HOSPITAL UNIVERSITI SAINS MALAYSIA FROM JANUARY 1991 TO DECEMBER 2021

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Introduction

- Paediatric Surgery services have been available in Hospital Universiti Sains Malaysia (HUSM) since 1989.
- Esophageal Atresia and Tracheoesophageal fistula (EA/TOF) is one of the congenital anomalies with estimated incidence of 1 in 2500 - 3000 live births.
- Death in EA/TOF rarely occurs nowadays, but if present is always secondary to other associated anomalies, prematurity or low birth weight.

Methodology & Objective

- This is a retrospective study of patients with EA and/or TOF admitted to HUSM Kubang Kerian from January 1991 to December 2021.
- List of patient derived from the Record Office's computer database and operative record books.
- All patients listed with EA and/or TOF who underwent surgical operation were included in the study and the data analyzed statistically by conducting the descriptive analysis.
- The aim is to study the clinical profile of the cases EA and/or TOF and various factors that influence the surgical outcome.

Results

- Total of 75 patients underwent surgical intervention for EA/TOF repair.

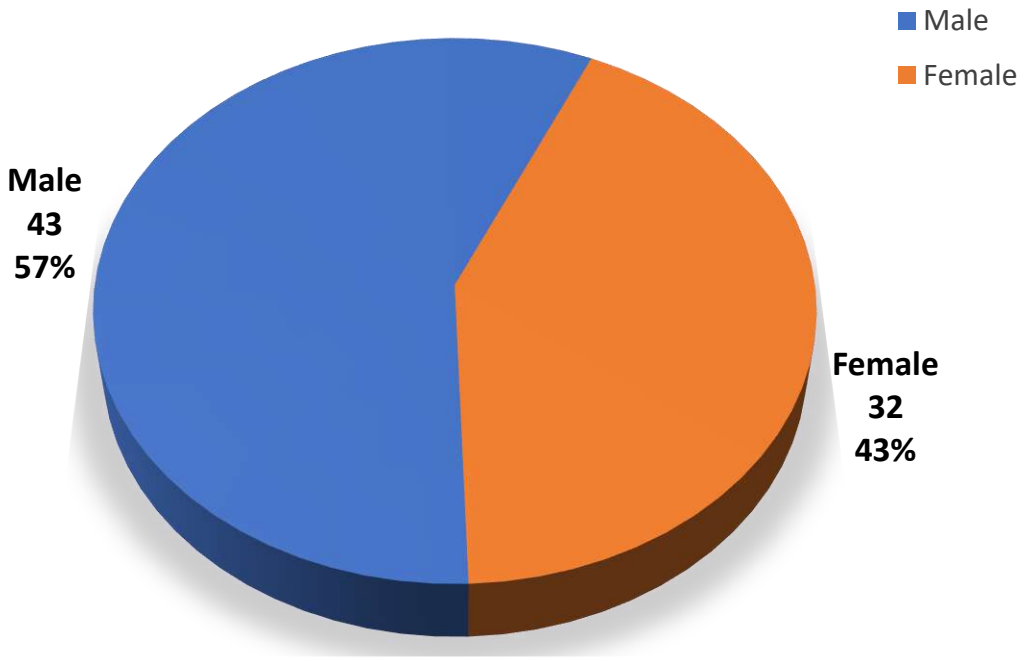


Figure 1: Sex distribution of EA/TOF patients

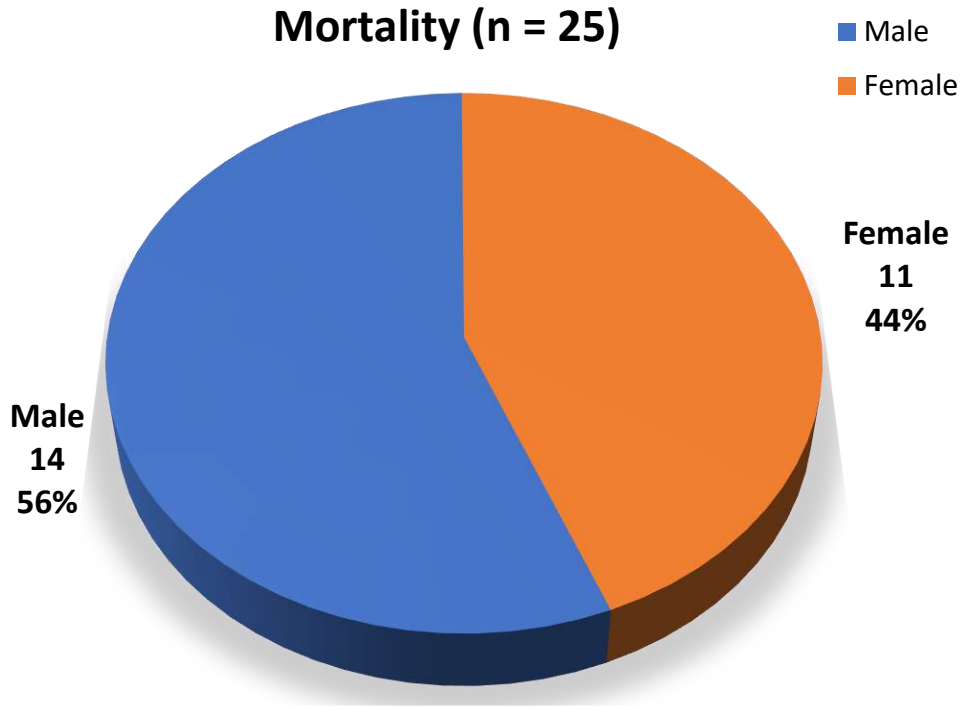


Figure 2: Sex distribution of mortality in EA/TOF patients

- Race
  - All cases were Malays (n = 75)
- Gestational age

GESTATIONAL AGE (WEEKS)	FREQUENCY	MORTALITY
≤ 37	20	11 (55.0%)
> 37	55	14 (25.4%)

Table 1 : Gestational age distribution and mortality for EA/TOF patients

- Birth weight

Table 2 : Birth weight distribution and mortality for EA/TOF patients

BIRTH WEIGHT (KG)	FREQUENCY	MORTALITY
< 1.8	14	9 (64.3%)
1.8 - 2.5	23	9 (39.1%)
> 2.5	38	7 (18.4%)

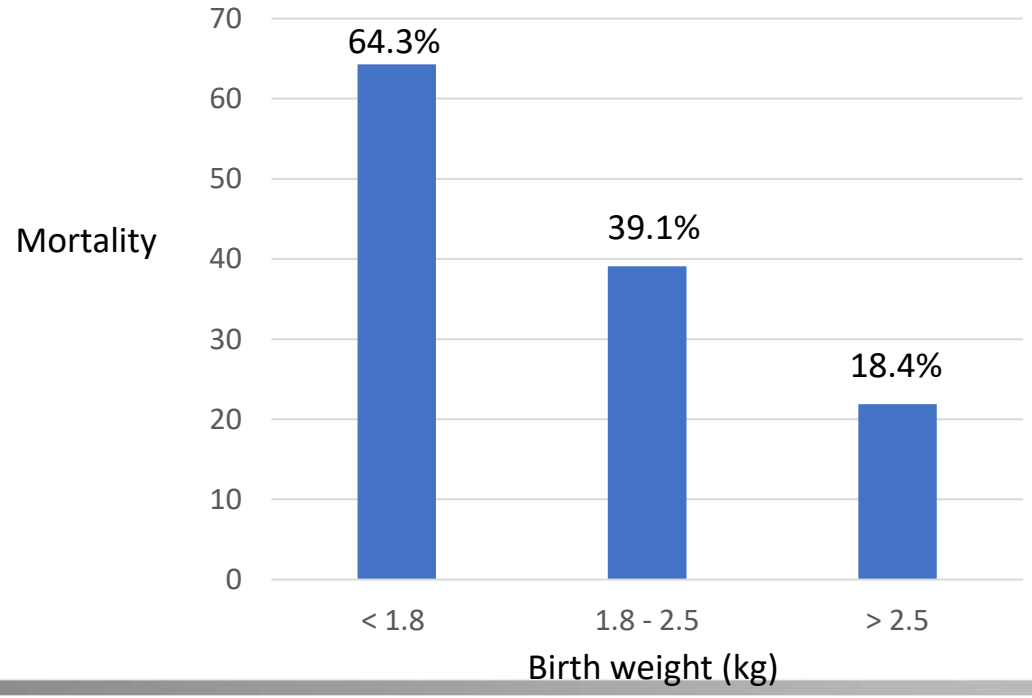


Figure 3:Mortality risk according to birth weight for EA/TOF

- Associated anomalies

No of anomalies	Frequency	Percentage (%)
0	23	31
1	26	35
2	15	20
3	4	5
4	5	6
5	1	1
6	1	1

Table 3 : Frequency of anomalies of EA/TOF patients

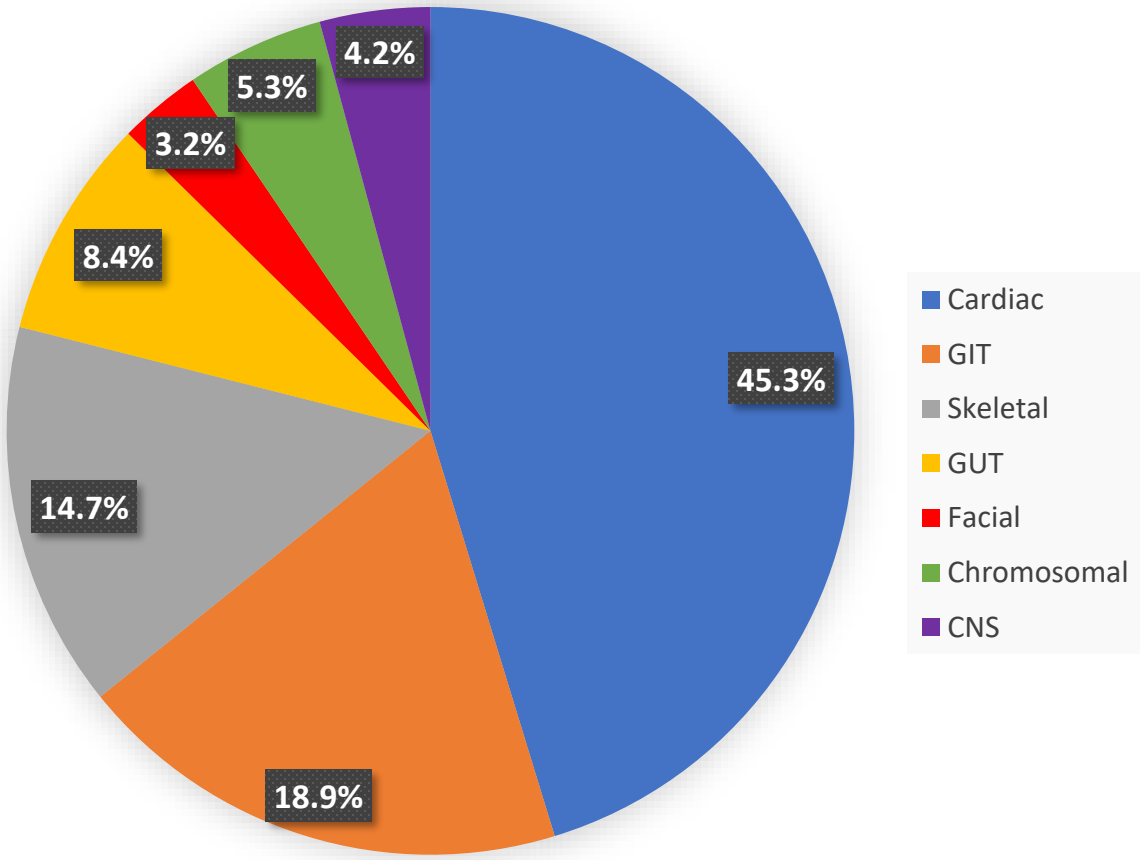


Figure 4: Percentage of congenital anomalies in EA/TOF

Congenital abnormalities	Frequency	Percentage
Cardiac anomalies	**43**	**45.3%**
Patent ductus arteriosus	19	
Ventricular septal defect	12	
Atrial septal defect	7	
Dextrocardia	3	
Teratology of Fallot	2	
Gastrointestinal anomalies	**18**	**18.9%**
Imperforated anus	16	
Duodenal atresia	2	
Skeletal anomalies	**14**	**14.7%**
Radius deformity	5	
Hemivertebrae	6	
Congenital Talipes Equinovarus	2	
Polydactyly	1	
Genitourinary anomalies	**8**	**8.4%**
Hypospadias	3	
Undescended testis	4	
Unilateral dysplastic kidney	1	
Facial abnormalities	**4**	**3.2%**
Cleft lip	3	
Cleft palate	1	
Chromosomal anomaly	**5**	**5.3%**
Down syndrome (trisomy 21)	5	
Central nervous system anomaly	**4**	**4.2%**
Hydrocephalus	3	
Corpus callosum dysgenesis	1	

Table 4 : Details on congenital anomalies of EA/TOF patients

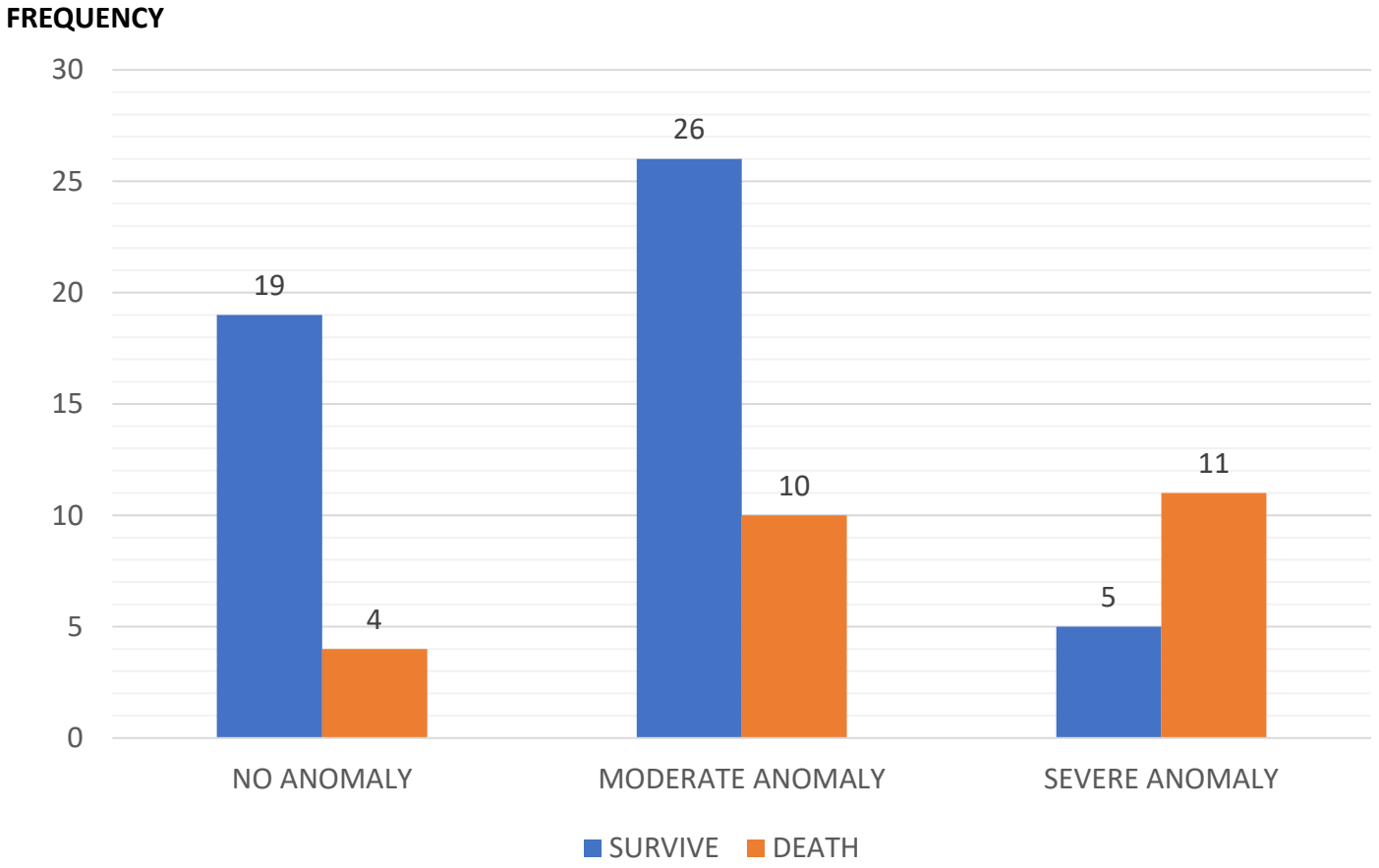


Figure 5: Distribution of patient and number of mortality according to the severity of associated anomaly (Waterston classification)

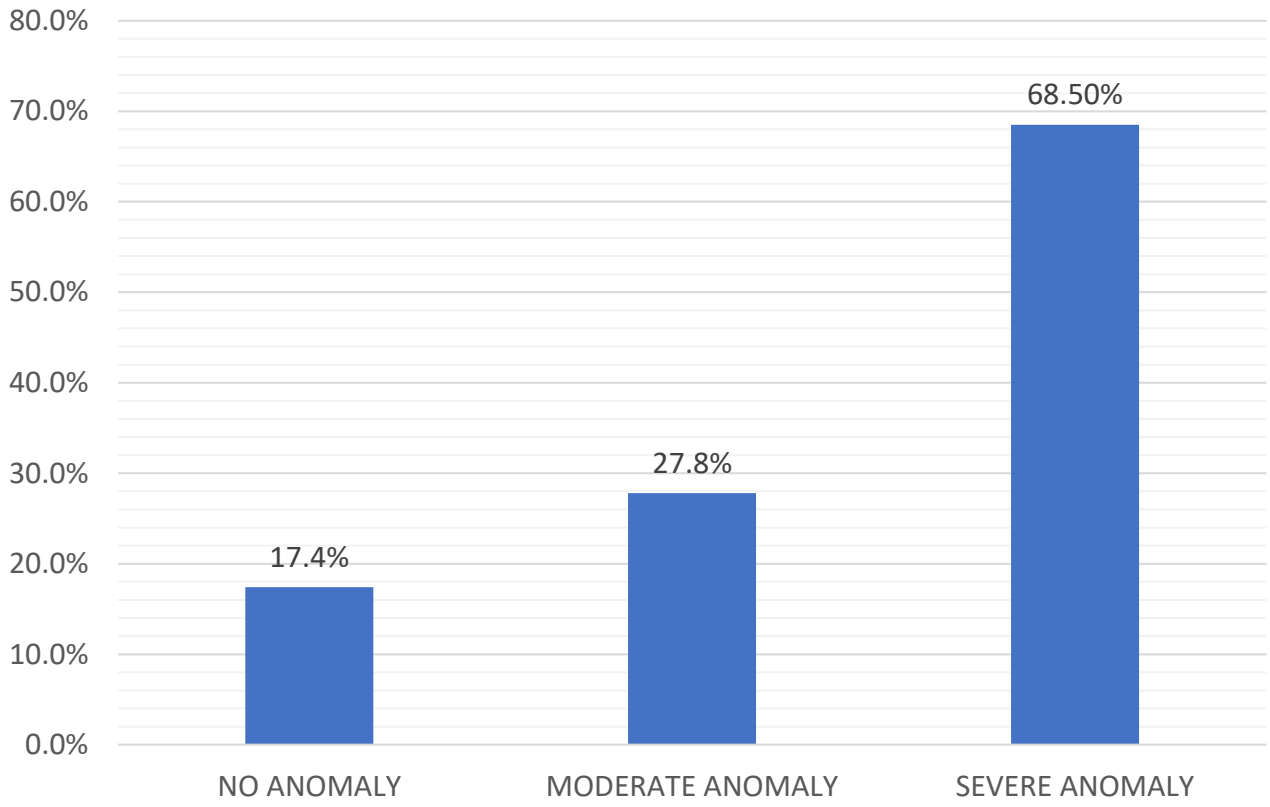


Figure 6: Mortality risk of EA/TOF patients with associated anomaly

- Aspiration Pneumonia and Ventilation prior to operation

ASPIRATION PNEUMONIA		VENTILATION		SEVERITY		MORTALITY	
YES	35 (46.7%)	YES	15	SEVERE	15	DIED	8
						ALIVE	7
		NO	20	SEVERE	6	DIED	3
						ALIVE	3
				MODERATE	14	DIED	4
						ALIVE	10
NO	40 (53.3%)					DIED	10
						ALIVE	30

Table 5: EA/TOF patient with aspiration pneumonia classified according to severity and number or mortality

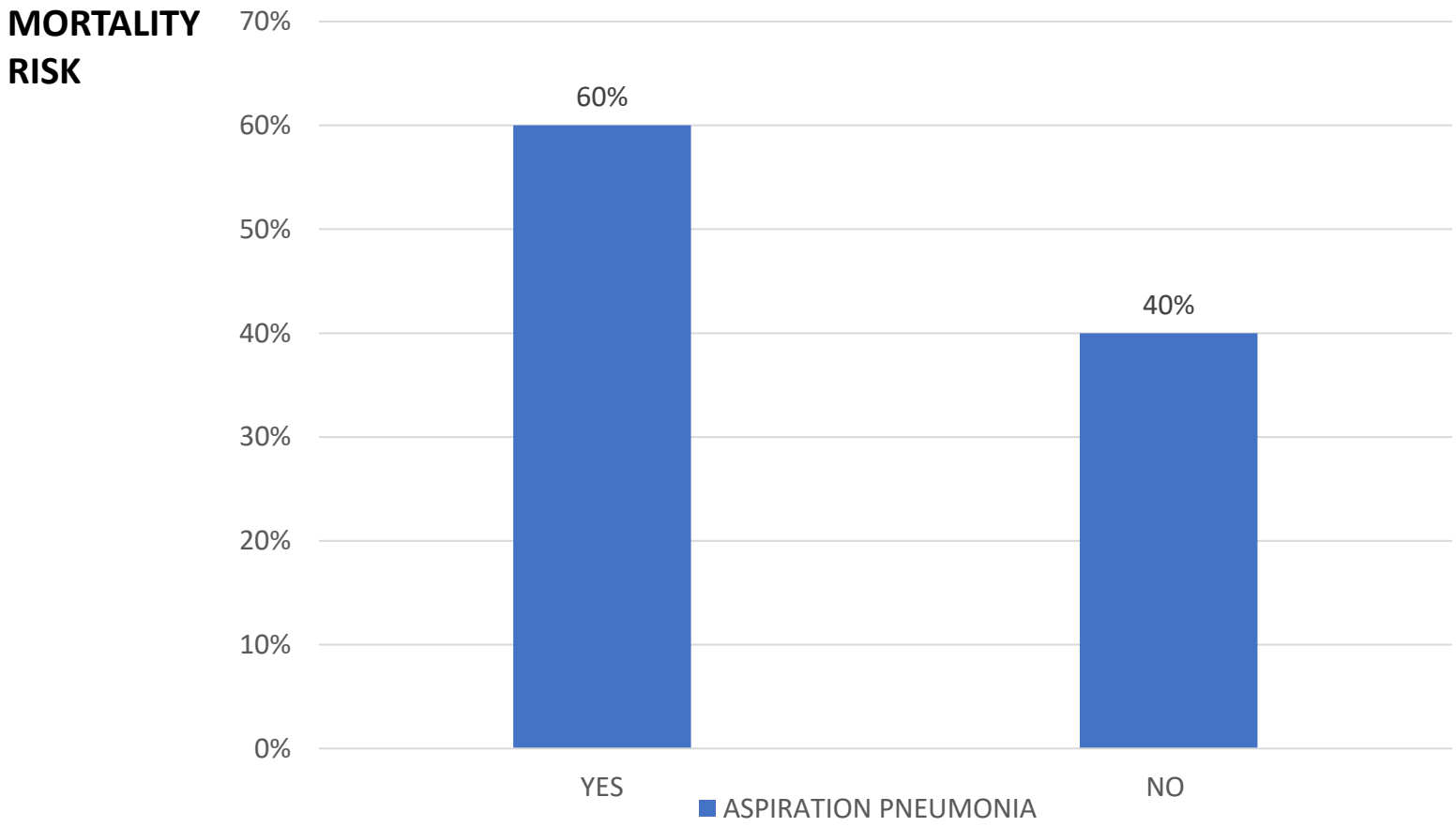
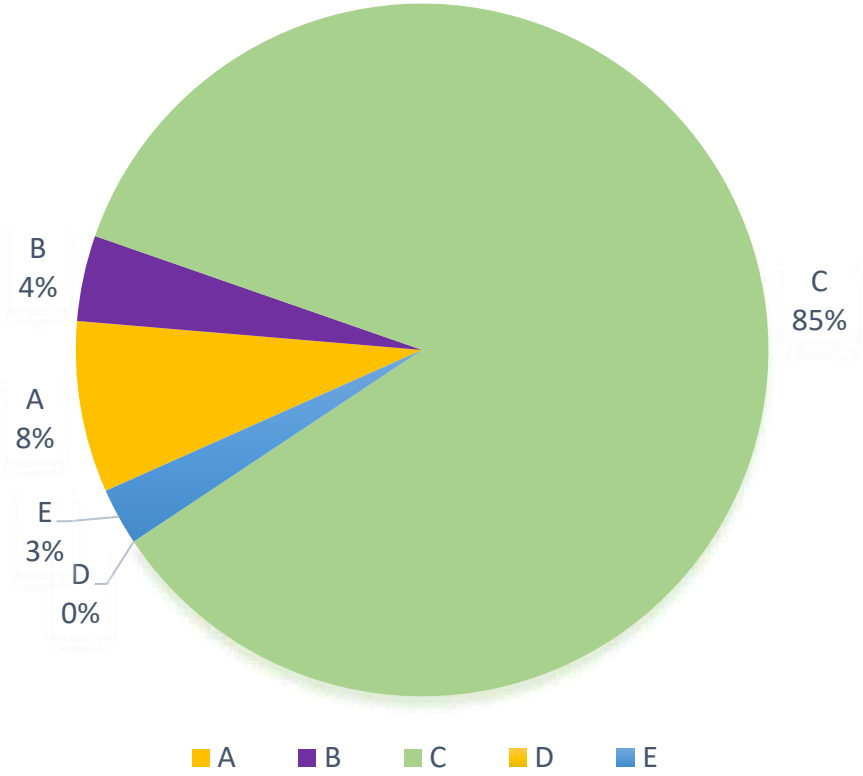


Figure 7: Mortality risk in patient EA/TOF with aspiration pneumonia prior to operation

- Type of Esophageal Atresia / Tracheoesophageal fistula based on Gross classification



A: Isolated Esophageal Atresia
B: Atresia with proximal fistula
C: Atresia with distal fistula
D: Atresia with double fistula
E: Isolated fistula (H type)

Figure 8: Percentage on each type of EA/TOF based on Gross classification

- Gap length

GAP LENGTH	FREQUENCY	MORTALITY
< 2cm	49 (67.1 %)	10 (20.4%)
> 2cm	24 (32.9%)	15 (62.5%)

Table 6: Gap length and mortality risk in EA/TOF patients

- Postoperative Complications

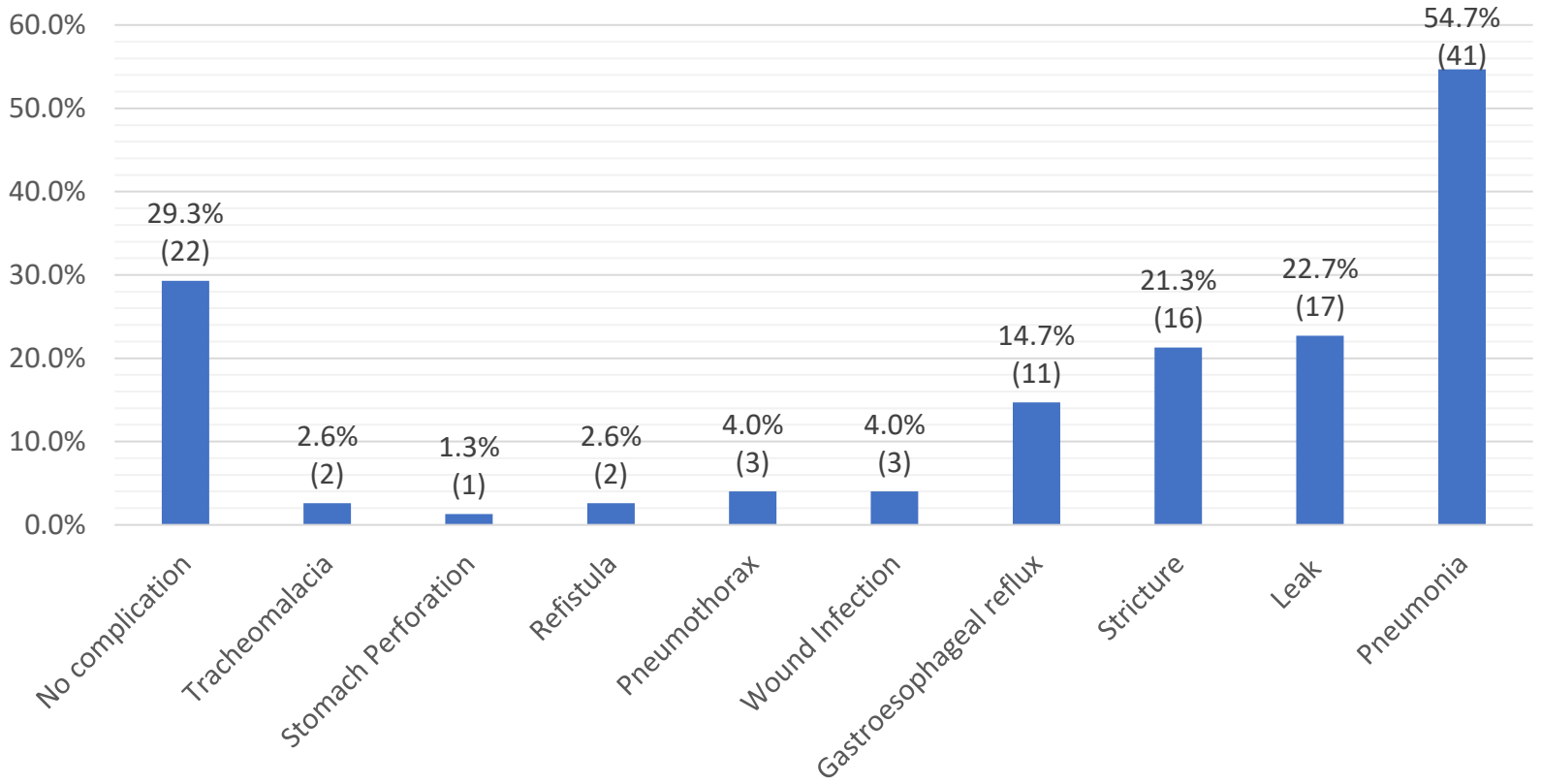


Figure 9: Postoperative complication of EA/TOF patients

- Mortality
  - Total of 25 death in this study (mortality rate 33.3%)
  - 50 patients (66.7%) survived with or without complication.
- Waterston classification

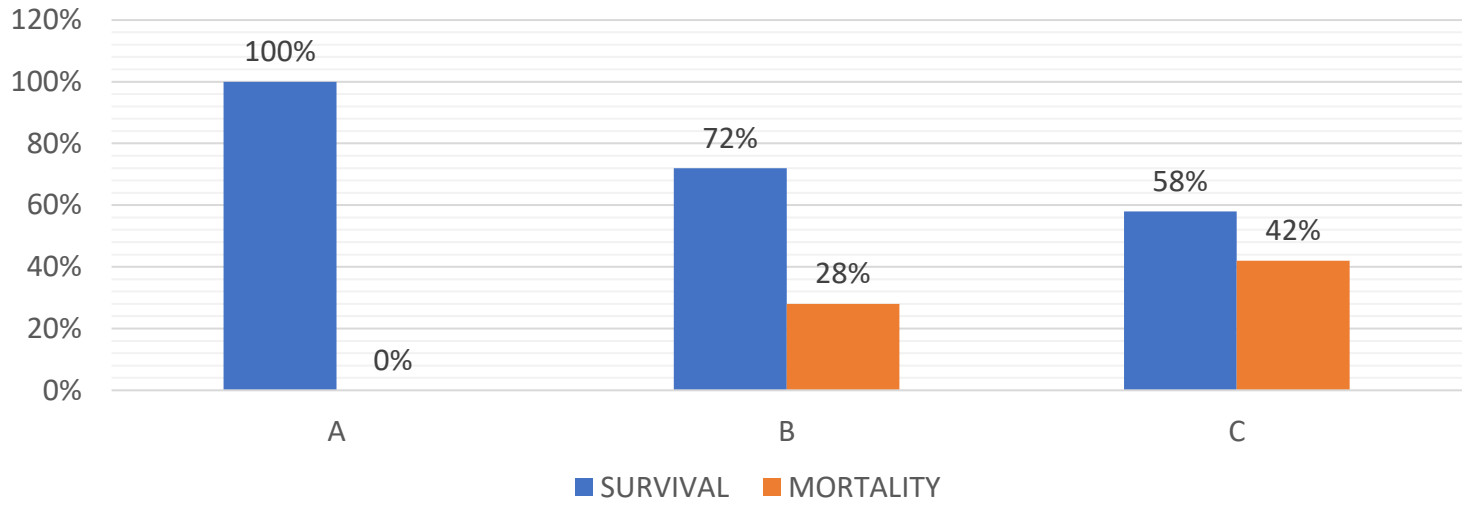


Figure 10 : Survival rate in EA/TOF patients according to Waterston classification

Discussion

- There was a male predominance (1.3 : 1) in our study. Cozzi *et al* and Niramis *et al* reported similar finding in their study.

- 26.7% of the infants were born prematurely, much lower compared to 39% noted by Myers et al. Mortality risk In premature infant is 55%, lower compared to 74% in Cozzi *et al*.

- In our series, low birth weight infant of < 1.8 kg had high mortality rate (64.3%) which is higher compared to Augusto Zani *et al* (50%).

- There were 52 infant (69%) with associated anomaly, higher compared to Niramis *et al* (65.2%). However, Yang *et al* reported higher incidence of associated anomaly (80%). The most common anomalies in this study is cardiac anomalies (45.3%), slightly higher compared to Encinas *et al* (42%).

- 84% of mortality in this study is associated with congenital anomalies, which is higher compared to *Davari et al* (74%).

- Preoperative aspiration pneumonia occurred in 46.7% of patients with 60% mortality rate. Louhimo *et al* reported slightly lower incidence of aspiration pneumonia (40%).
- In most study, Type C Esophageal Atresia (Gross classification) is the commonest one. In our centre, 85% of the cases is Type C followed by Type A.
- High mortality rate in patient with atretic gap > 2cm (62%). Similar finding was noted by Hands *et al.*
- The overall mortality rate in this series was 33.3%.

WATERSTON CLASSIFICATION	Konkin et al, 2000 (Canada)	Narasimman et al , 2009 (HSB, Kedah)	Niramis et al, 2013 (Thailand)	Kamal et al, 2015 (India)	HUSM 2021 (Kelantan)
GROUP A	100%	100%	100%	98%	100%
GROUP B	100%	89%	91.5%	59%	72%
GROUP C	80%	33%	48.8%	15%	58%

Table 8: Comparison of survival rate of patients with EA/TOF according to Waterston classification

Conclusion

- We found birth weight, severity of associated anomaly, preoperative aspiration pneumonia and atretic gap length are significantly influence the outcome of patients with EA/TOF in HUSM.
- Based on Waterston classification, patient without other anomalies has a 100% chance of survival.
- Our limitations for this is study is due to its retrospective nature and dependence upon the data that had been collected by the previous surgeon. We are unable to retrieve the raw data from the previous study to proceed with further statistical analysis.
- For a better association and correlation, a large sample size is needed. This study should be continued and reviewed every 5 years.

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