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Esophageal Atresia and Tracheoesophageal Fistula, is Waterston`s Classification Still Valid?

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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ABSTRACT

Aim: Due to advances in the operative techniques and neonatal care, the Waterston's system of prognostic classification for oesophageal atresia (EA) is no longer followed in the developed-world.
Objective: The aim of this study was to evaluate the validity of risk stratification according to the Waterston`s Classification system for the repair of EA/TEF in the developing-countries.
Methods: This is a retrospective study, in which all 30 cases of congenital EA/TEF admitted to Bolan Medical-College Hospital, Quetta from July 2013 to January 2018 were studied. Risk-stratification was done according to the Waterston's classification system. Institutional review board of bolan medical college authorized the study.
Results: Among the 30 records reviewed in this study, 90% of the patients were born outside BMCH. The diagnosis was made postnatally by means of nasogastric tube and chest X-ray (83%). The most common type of TEF was Type C (98%). The most common post-operative complication was pneumonia (43%). Overall survival rate was 83%. Cardiac-anomalies were the major associated cause of mortality. All patients who died had major cardiac anomalies p -<0.05.
Risk stratification according to Waterston's-classification system showed 100% survival in Groups A, 82% in Group B and 75 % in Group C.

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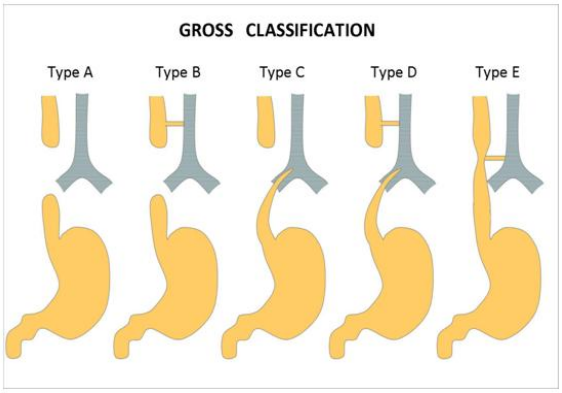
Conclusion: We conclude that the risk stratification according to Waterston's-classification, which is now being considered obsolete in the developed world, is still valid in the developing countries like Pakistan because it is a relevant prognostic indicator it helps in deciding the surgical intervention, and has better correlation with survival than the other factors.

Keywords: Oesophagal atresia; tracheoesophageal fistula; Waterston`s classification.

1. INTRODUCTION

Oesophagal atresia refers to a congenitally interrupted oesophagus in which the upper end of the oesophagus ends in a blind-ended pouch rather than connecting to the stomach. A tracheoesophageal fistula (TEF) is an abnormal connection between the trachea and the oesophagus. The lack of oesophagal patency prevents swallowing which causes drooling of copious amounts of saliva in the neonates. It also causes aspiration of saliva or milk leading to aspiration pneumonitis. The lungs of the babies born with EA and distal TEF are exposed to gastric secretions. Air from the trachea may also pass down the distal TEF leading to a dangerous condition known as acute gastric perforation. Robert E. Gross described the most accurate classification system for oesophagal atresia [1] (as seen in the Image below):



According to his classification, the types of oesophagal atresia are as follows:

- Type A - Oesophageal atresia without fistula. It is also called pure oesophageal atresia (10%)
- Type B - Oesophageal atresia with proximal TEF (<1%)
- Type C - Oesophageal atresia with distal TEF (85%)
- Type D - Oesophageal atresia with proximal and distal TEFs (<1%)
- Type E - TEF without ooesophagal atresia (also called H-type fistula) (4%)

No positive correlation has been found between teratogens and oesophagal atresia. At present, there is a strong consensus among the scientific community that oesophagal atresia has no genetic basis [2]. However, tracheoesophageal fistula has been found to be caused as a result of embryopathologic process in many studies [2-4].

The incidence of oesophagal atresia is 1 case per 3000-4500 births [5,6] and this frequency has been found to be decreasing for unknown reasons [7].

The first prognostic classification for oesophagal atresia was proposed by David J. Waterston et al. in 1962 (Table 1) who identified the importance of birth weight, pulmonary status and a number of other congenital anomalies as factors influencing neonatal survival [8]. However, since then a number of investigators have questioned the validity of this system of classification [4,6,7,9].

The aim of our study is to determine the validity of Waterston's system of classification in developing countries like Pakistan.

Table 1. Waterson's classification

Waterson's classification		
Category	**Weight/ co-morbidities**	**Surgical timing**
Category A	>2500 grams	Can undergo surgery
Category B	1800 – 2500 g, pneumonia or congenital anomaly	Short term delay, needs stabilizing treatment prior to surgery
Category C	<1800 g or severe pneumonia or congenital anomaly.	Required staged repair.

2. MATERIALS AND METHODS

This is a retrospective study where the case records of all patients admitted to BMCH, Quetta with a diagnosis of congenital EA/TEF from July 2013 to January 2018 were reviewed (Table 2). Institutional Review Board of Bolan Medical College authorized the study. Several parameters were taken into consideration including presenting symptoms, associated cardiac and other major congenital anomalies, and the modalities that were used to confirm the diagnosis.

We also studied the different types of tracheoesophageal fistulas in the patients, the type of corrective surgery performed, and the associated early and late complications of the procedure. Risk stratification was then carried out using the Waterston's Classification system and mortality in each Group A, B and C was computed.

The data was analysed using SPSS VERSION-20.

Table 2. Oesophagal atresia and tracheoesophageal fistula: Cases reported at the Bolan Medical College Hospital, Quetta (2013-2018)

Total Number of patients	30
Overall survival rate	83%
Overall mortality	16.6%

3. RESULTS

A total of 30 cases of EA/TEF received at Bolan Medical College Hospital, Quetta were studied. Among those, 27 (90%) of the cases were delivered outside BMCH, Quetta.

3.1 Diagnosis

The antenatal diagnosis was suspected in 9 cases, where polyhydramnios (an abnormally high level of amniotic fluid) was found in 8, and the absence of the gastric bubble in 1 case. In the postnatal period, the diagnoses was confirmed in all patients using chest X-ray with N/G tube in 25 (83%), contrast studies in 4 (13%), and bronchoscopy in 1 (4%).

3.2 Presenting Symptoms

The total percentages of symptoms among patients were as follows:

1. Excessive salivation (80%)
2. Vomiting (65%)
3. Cough (60%)
4. Choking (65%)
5. Respiratory distress (47%)
6. Cyanotic spell (45%)
7. Regurgitation (40%)

3.3 Associated Anomalies

Two-thirds (20) of the patients had associated anomalies. The most common being cardiac (8), followed by renal (3), GIT (3), and others (6) (Table 3).

Table 3. Associated anomalies presented with EA/TEF

Associated anomalies	Number of patients n=20
Cardiac	**8**
Tetralogy of Fallot	3
VSD	4
ASD	1
Renal	**3**
Right atrophic kidney	1
Left absent kidney	1
Right hydronephrosis	1
Other anomalies	**7**
Down`s syndrome	1
Pierre Robin Syndrome	1
Hydrocephalus	1
Hypospadias	1
Talipes Equinovarus	1
VACTERL	1
Polydactyl	1

3.4 Operative Management

Preoperatively, 10 (33%) patients had pneumonia, 18 (60%) required admission in the Neonatal intensive care unit and 9 (30%) patients required ventilation preoperatively.

The minimum age at surgery was 6 hours and maximum being 21 days (mean=6.5 days). Primary TEF repair was performed in 12 patients, gastrostomy + primary TEF repair in 9 patients and Gastrostomy and delayed TEF repair in 9 patients. The mean hospital stay was 14 days, with a mean NICU stay of 10 days. 50% of patients required ventilation postoperatively.

Among the 30 patients, the most common type of tracheoesophageal fistula was Type C (98%). One of the patients only had oesophagal atresia, and one patient had the H-type Tracheoeso-phageal fistula.

3.5 Complications of Surgery

Among the 30 patients, 20 (66.6%) had early complications, the most common being sepsis with pneumonia in 13 (43%). Others complications included anastomotic leakage 3 (10%), wound infection in 2 (6.6%), aspiration pneumonia in 1 (3%) and pneumothorax in 1 (3%) patient.

Sixteen of the patients experienced late complications, the most common being regurgitation in 6 (20%) patients. Other late complications included dyspnea in 2 (6.6%), esophageal stricture in 2 (6.6%), gastro-esophageal reflux in 2 (6.6%), strictures in 2 (6.6%), tracheomalacia in 1 (3%) and hiatus hernia in 1 (3%).

3.6 Risk Stratification and Survival

Risk stratification was performed according to the Waterston's system of classification and then survival in each group was computed (Table 4). Among the 30 patients, 4 (13.3%) belonged to Group A, 14 (46.7%) to Group B, and 12 (43%) to Group C. The mortality in Group A was 0%, Group B 2 (6.5%) and Group C 3 (10.5%).

4. DISCUSSION

Waterston`s Classification system is used to define risk categories for patients with oesophagal atresia and tracheoesophageal fistula. The risk stratification is based on birth weights, gestational age, pulmonary status and the presence of associated congenital anomalies is critical in identifying high-risk infants and their associated poor outcome. In this study, we tried to evaluate the validity of Waterston`s Classification system for the repair of EA/TEF in the developing countries.

Since the first successful repair of oesophagal atresia and tracheoesophageal fistula (EA&TEF) in 1941, the outcomes have improved remarkably [10,11]. Medical advancements such as improved neonatal care, earlier recognition and management of associated congenital anomalies, improvement of neonatal anaesthesia and NICU monitoring and refinement of surgical techniques principally are responsible for the increased survival rate of the patients with EA/TEF. Waterston's prognostic classification of oesophagal atresia has been used by healthcare professionals for decades around the world. A great number of technical advances, mainly in neonatal intensive care units have led to a significant reduction in mortality. Although the Waterston classification continues to be widely used but increased survival in the highest risk groups in this classification has led to new classifications being described such as the Montreal system which identifies more accurately the children at highest risk than the Waterston classification [11,12].

In the developing countries delayed diagnosis of congenital anomalies associated with EA/TEF, poor transport facilities and inaccessible healthcare facilities and lack of neonatal care and operative techniques lead to increased morbidity and mortality [13].

In this study, we evaluated the infants who presented with a diagnosis of EA & TEF retrospectively (from July 2013 to January 2018) on the basis of Waterston`s classification. Two-thirds of our patients had various associated congenital anomalies among which cardiac anomalies were the most common (40%). Preoperatively, 33% (10 patients) had pneumonia, 60% (18) were admitted directly in the NICU and 30% (nine) required preoperative ventilation. Postoperatively, 50% of the patients were kept on a ventilator. Twelve patients had primary repair of EA and TEF without gastrostomy. These were the patients with no respiratory complications. Nine patients underwent gastrostomy with TEF repair and nine patients first underwent Stamm gastrostomy and delayed TEF repair. These patients had major cardiac anomalies.

Table 4. Survival in different groups according to the Waterston's classification in patients of EA/TEF at Bolan Medical College Hospital, Quetta (N=28)

		% of patients		Survival rate
Group A	>2.5 kg birth weight	4 patients	(13.3%)	100%
Group B	1.8-2.5 kg birth weight >2.5 kg birth weight with moderate congenital anomalies and pneumonia	14 patients	(46.7%)	82%
Group C	<1.8 kg birth weight Several congenital anomalies	12 patients (43%)		75%

Type C (EA with distal tracheoesophageal fistula) was the commonest variety (93%). Early complications included an anastomotic leak in three patients (10%), pneumonia in thirteen patients (43%), and wound infection in 2 (7%). Sixteen patients experienced late complications, which include regurgitation in six patients (20%), GER in two patients (7%), oesophagal stricture requiring dilatation in two patients (7%), tracheomalacia in one patient and hiatus hernia in one patient.

Cardiac anomalies associated with EA/TEF have been identified as a poor prognostic factor and a frequent cause of death [9,14]. The mortality rate is highest in patients with congenital cyanotic heart disease; the reported incidence is (40-50%). This warrants the need for Echo in every patient of EA/TEF. In our study, all patients who had congenital heart disease died after hospitalization.

Risk stratification according to the Waterston classification showed 100% survival in Groups A, 82% in Group B and 75 % in Group C. We have found that Waterston's prognostic criteria is good for evaluating prognosis in the neonates suffering from EA/TEF in the developing countries which is consistent with the studies of B Eradi et al. [15].

5. CONCLUSION

We conclude that the risk stratification according to Waterston's classification, which is now being considered obsolete in the developed world due to advances in neonatal care and operative techniques, is still valid in the developing countries because it is a relevant prognostic indicator, it helps in deciding the surgical intervention, and has better correlation with survival. This study had limitations due to small sample size.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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