

Thirteen Ribs and Long Gap Oesophageal Atresia: The Embryological Hypothesis for Exploration

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Abstract

Context: Since the gap between two atretic segments of oesophagus is a critical determinant of prognosis for oesophageal atresia/tracheoesophageal fistula (EA/TEF), the search for a surrogate non-invasive pre-operative marker of long gap atresia continues. **Aims:** The purpose of the study was to compare the presence of normal and supernumerary ribs with length of EA and survival rates. **Settings and Design:** A prospective observational study was conducted at a tertiary care referral neonatal intensive care unit in North Karnataka, India, from January 2016 to June 2019. **Subjects and Methods:** Amongst babies with EA/TEF, pre-operative radiograph helped determine the number of ribs, and babies were divided into two groups; Group I: babies with 12 ribs and Group II: babies with supernumerary ribs. **Statistical Analysis Used:** Nominal variables were expressed as percentage and continuous variables as mean standard deviation. MedCalc software was used to compare proportions and means. A $P < 0.05$ was considered statistically significant. **Results:** Of the 61 cases, 51 were operated. Long gap EA was predominantly seen amongst babies in Group II (40% in Group II vs. 27% in Group I, $P = 0.424$). Survival rates by percentage were lower in babies in Group II (60% in Group II vs. 80% in Group I, $P = 0.188$). Both the above findings were proven statistically insignificant. The overall survival rate amongst the study population was 78.4% (39/51). **Conclusions:** Supernumerary ribs were associated with a higher occurrence of long gap EA and lower survival rates, though statistically insignificant. Multicentre collaboration may provide significant input for strengthening or refuting the above hypothesis.

Keywords: Fistula, long gap oesophageal atresia, supernumerary ribs, thirteen ribs, tracheoesophageal

INTRODUCTION

The search for non-invasive markers of long gap oesophageal atresia (EA) continues as they can serve as important predictors for type of surgery as well as prognosis of the baby. The fascinating association between 13 ribs and long gap EA was first ever conceived by a paediatric surgeon from Mumbai, India.^[1] It was hypothesised that the increasing number of ribs in thorax increased the gap between the two atretic segments of oesophagus. Since then, the literature has added on from paediatric surgeons worldwide to test the hypothesis further. The present article focuses on the same concept and adds on to the existing scarce literature. The article compares the prevalence of long gap atresia amongst babies with normal and supernumerary ribs and also highlights the survival rates amongst the two groups of babies.

SUBJECTS AND METHODS

A prospective observational study was conducted from January 2016 to April 2019 in northern Karnataka, India. The neonatal unit is a tertiary care referral unit that cares for newborn babies from all over the district as well as neighbouring districts. Demographic data of mother–baby dyad were collected. All the babies with tracheoesophageal fistula (TEF) who were admitted to neonatal intensive care unit were included in the study. A total of 61 consecutive admissions were eligible for the study over a period of 3 years. Those babies who did not undergo surgery were excluded from the study.

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The study population was divided into two groups based on the number of ribs on the chest radiograph (anteroposterior view), which was taken at admission [Figure 1]. The first group (Group I) consisted of babies with 12 ribs and the second group (Group II) consisted of babies with 13 and more (supernumerary) ribs. The primary objective of the study was to compare the prevalence of long gap EA amongst the two groups. The secondary objective was to compare the survival rates amongst the two groups. Along with TEF, other associated anomalies were recorded before surgery. If there was association of EA with other anomalies, parents were further counselled regarding the short- and long-term implications of the multiple morbidities. After taking informed consent from parents, baby was posted for surgery. Intra-operative findings were carefully noted.

There is no consensus about the definition of long gap EA with different studies using varying definitions. The first study from Mumbai, India, defined long gap as distance of >2 vertebral bodies between the atretic segments.^[1] The Australian experience published in 2008 depicted a survey amongst paediatric surgeons. Forty per cent of Australian surgeons defined long gap as a gap more than 3–4 vertebral bodies and 24% considered the absence of TEF as an indicator of long gap.^[2] According to a study protocol registered with *ClinicalTrials.gov*, a gap of >3.5 cm has been defined as long gap EA.^[3] Borrowing from the above studies, the present study defines long gap atresia as a gap of >3.5 cm. Figure 2 (a – atretic segment of oesophagus and b - fistula tract) shows an intraoperative picture of small gap atresia in a baby with supernumerary ribs. Post-operatively, clinical course of these babies was recorded. Other morbidities such as septicaemia and anastomotic leak were also noted.

Statistical methods

The nominal variables were expressed as percentages. Comparison of proportions amongst the two groups was done using the Chi-square test. When the number in any cell had a value < 5, Fischer's exact test was used to compare proportions. Continuous variables were expressed as mean \pm standard

deviation. Student's *t*-test and Mann–Whitney *U*-test were used for skewed and normally distributed variables, respectively. The *P* values were confirmed with MedCalc software. A *P* < 0.05 was considered statistically significant.

RESULTS

A total of 61 EA cases were admitted to Shanti Hospital, Bagalkot, a tertiary care referral centre in North Karnataka, India, from January 2016 to April 2019. Of the total study population, 48 babies had 12 ribs and 12 babies had 13 ribs on chest radiograph. One baby had 14 ribs on chest radiograph in association with EA.

Of the 48 babies with 12 ribs on chest radiograph, 41 babies were operated. Of the 12 babies with 13 ribs on chest radiograph, nine were operated. Seven babies in the former group and three babies in the latter group were not operated as they suffered from multiple congenital anomalies and parents were not willing for surgery, and hence, these babies were excluded from analysis. One baby with 14 ribs was also operated upon and included in Group II [Flow chart 1].

As evident from Table 1, the average maternal age was 23.6 and 24.5 years in the first and second groups, respectively. The native areas from where the cases hailed were about 71 km and 77 km of radius in and around Bagalkot, respectively. The average birth weight amongst the two groups was 2395 \pm 579 g and 2093 \pm 448 g, respectively. Premature babies constituted 35% (18/51) of the study population. Thus, there was no statistically significant difference amongst the two groups with respect to baseline characteristics of the study population.

Table 2 describes the survival rates and compares the association of long gap EA with the number of ribs (primary and secondary outcomes of the study). Long gap EA was found in 27% (11/41) and 40% (4/10) in Group I and Group II, respectively. Although the occurrence of long gap atresia was higher in supernumerary ribs group, the comparison was not



Figure 1: Chest radiograph showing 13 ribs and oesophageal atresia



Figure 2: Intra-operative picture showing mild gap oesophageal atresia in the same baby with 13 ribs. a – atretic segment of oesophagus, b – fistula tract

statistically significant ($P = 0.424$). Survival rates were lower in Group II compared to Group I (80% [33/41] in Group II vs. 60% [6/10]). It is to be noted that the total number of babies in Group II is lower ($n = 10$) and with every mortality, the survival percentage comes down drastically. The overall survival rate amongst the study population is 78.4% (40/51).

There were other comorbidities contributing to the mortality rates amongst both the groups. Associated anomalies were found in 37% (15/41) and 30% (3/10) of the babies in Group I and Group II, respectively.

Table 3 describes the comorbidities present amongst the babies in both the study groups. The most common associated anomaly was cardiac malformation found in 27% (14/51) of the babies included. Cardiac malformations were higher by two folds in Group II (supernumerary ribs), almost reaching the statistical significance ([22% [9/41] in Group I vs. 40% [4/10] in Group II, $P = 0.08$).

Table 4 describes the characteristics of babies who died in both the subgroups. There were a total of 12 deaths in the study cohort, of whom eight babies were in Group I and four babies were in Group II.

DISCUSSION

Supernumerary ribs and long gap EA occurring in concurrence have attracted the attention of paediatric surgeons both from India and abroad. In 1997, Kulkarni *et al.* from Mumbai, India, had reported the significant association of 13 pairs of ribs

with long gap EA. A total of 61 cases of EA were studied, of whom nine patients had 13 ribs and each one of them had a long gap atresia.^[1] Kulkarni *et al.* had proposed that presence of supernumerary ribs indicates hypersomatization at thoracic level causing increased length of developing thorax and placing the foregut under stretch, thus causing long gap atresia. Subsequently, other cohorts of EA and the co-occurrence of 13 ribs were reported in the literature, and the association with long gap atresia was studied further.

Bosenberg and Hadley^[4] reported eight cases of 13 ribs and none of the babies in the cohort had long gap EA. Further, in the same study, there were 14 babies with long gap atresia and none of them had 13 ribs. Another study by Durell *et al.* reported a cohort of ten babies with 13 ribs. This study from Leicester, UK, found that none of the babies with extra mesodermal segment had long gap atresia. This study also had babies with 11 pairs of ribs and EA.^[5] Both the above studies were in contrast to previous report by Kulkarni *et al.*^[1]

The present study had 12 cases of EA in association with 13 ribs and nine of them were operated. Of the nine cases, four had long gap atresia. The baby with 14 ribs had mild gap EA. The presence of long gap EA amongst the babies with supernumerary ribs was definitely higher than babies with 12 ribs, though the association was not statistically significant ($P = 0.424$) [Table 2].

As it is evident from Table 5, the Indian subcontinent has higher prevalence of co-occurrence of 13 ribs with long gap

Table 1: The mother- infant characteristics that were studied

Characteristic	Group I (12 ribs) ($n=41$)	Group II (supernumerary ribs) ($n=10$)	<i>P</i>
Maternal age (years), mean±SD	23.6±3.1	24.5±4.2	0.45
Distance of native area (km), mean±SD	71±41	77±57	0.70
Consanguinity	26/41 (63)	6/10 (66)	0.86
Low birth weight	21/41 (51)	8/10 (80)	0.10
Prematurity	14/41 (34)	4/10 (40)	0.72
Average duration of hospital stay (days)	13.5±5.4	13.7±5.25	0.92

SD: Standard deviation

Table 2: The survival rates and compares the association of long gap oesophageal atresia with the number of ribs

Outcome	Group I (twelve ribs) ($n=41$), <i>n</i> (%)	Group II (supernumerary ribs) ($n=10$), <i>n</i> (%)	<i>P</i>
Survival rate	33/41 (80)	6/10 (60)	0.188
Long gap EA	11/41 (27)	4/10 (40)	0.424

EA: Oesophageal atresia

Table 3: Compares prevalence of risk factors/comorbidities amongst both the groups of babies

Co-morbidities/characteristics	Group I (twelve ribs) ($n=41$), <i>n</i> (%)	Group II (super-numerary ribs) ($n=10$), <i>n</i> (%)	<i>P</i>
Culture positive sepsis	9/41 (22)	4/10 (40)	0.25
Anastomotic leak	4/41 (10)	2/10 (20)	0.39
Cardiac defects	9/41 (22)	5/10 (50)	0.08
Anorectal malformation	4/41 (10)	0	-
Isolated TEF/EA	26/41 (63)	7/10 (70)	0.65

EA: Oesophageal atresia, TEF: Tracheoesophageal fistula

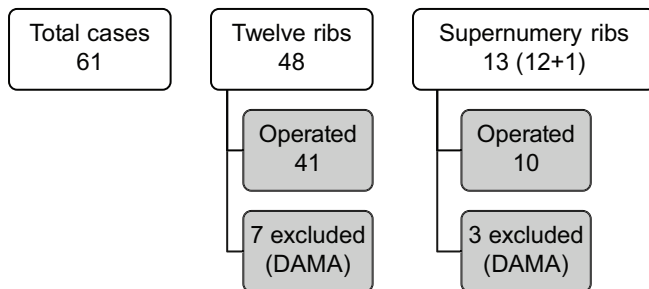
Table 4: The characteristics of babies who expired in the study cohort

Characteristic	Group I (twelve ribs) (n=8) n (%) / mean ± SD	Group II (supernumerary ribs) (n=4) n (%) / mean ± SD
Male gender	3 (38)	3 (75)
Maternal age (years)	22.1±2.7	23.25±3.2
Average birth weight (g)	2357±451	1825±330
Pre-maturity	4 (50)	3 (75)
Cardiac anomalies	1 (13)	3 (75)
Culture-positive sepsis	5 (63)	2 (50)
Wide gap	3 (38)	4 (100)

SD: Standard deviation

Table 5: Compares the present study with previous studies that have reported the association of 13 ribs with long gap oesophageal atresia

Name of the study	Year of the study	Total number of babies (n)	Number of babies with ≥13 ribs (n)	Long gap atresia and ≥13 ribs, n (%)
Kulkarni <i>et al.</i> 1997 ^[1]	1991- 1996	61	9	9 (100)
Bosenberg and Hadley 1998 ^[4]	1988- 1997	155	5	0
Durell <i>et al.</i> 2017 ^[5]	2005- 2012	76	10	0
Present study	2016- 2019	61	12*	4 (40) [#]

*Non-operated cases are included, [#]Of the ten operated cases, four had long gap atresia**Flow Chart 1:** Flow chart depicting distribution of study subjects based on number of ribs (DAMA: Discharge against medical advice)

EA. Two other studies by Bosenberg and Hadley^[4] and Durell *et al.*^[5] have shown that 13 ribs on chest radiograph were not associated with long gap atresia. This finding might have an epidemiological and genetic implication since contrasting evidence has emerged from countries other than India. This observation, however, needs additional evaluation.

In 1973, Band-Taylor *et al.* studied association between extra mesodermal segments and mortality. The study consisted of nine babies with 13 ribs. However, there was no mention about the occurrence of long gap atresia. The study consisted of 13 babies with vertebral anomalies and showed no association of musculoskeletal anomalies on mortality or prognosis of the study participants.^[6]

The strength of the present study is the total volume of cases with EA over a short period of 3 years, which may also carry epidemiological importance in a locality that has a higher prevalence of consanguineous marriages and congenital and chromosomal anomalies.

The limitation is the small number of cases with supernumerary ribs that limits the comparison between the two groups. The number is further truncated by cases which are not operated or discharged against medical advice. Every baby with TEF/EA could not be subjected to surgery considering the multiple comorbidities with cardiac anomalies being the predominant issue and further added on by the financial constraints of the population served by the hospital in a developing country.

In a purely statistical sense, it is prudent to say that the presence of supernumerary ribs was not associated with long gap EA in the present study. However, clinically 40% of the babies with 13 ribs had long gap EA [Tables 2 and 5]. Nevertheless, the findings of the present study partially provide important inputs for management and prognostication of cases of EA. To illustrate this point further, a pre-operative chest X-ray showing supernumerary ribs points towards a possibility of wide gap atresia. It can alert the surgeon to counsel the parents regarding the need for staged surgery if the primary anastomosis is not feasible.

Thus, the association between supernumerary ribs and long gap atresia needs to be studied over a long period of time and needs multicentre collaboration to increase our understanding of this interesting association. However, the study remains to be ongoing at our centre and the search for other markers of long gap atresia continues in time.

CONCLUSION

The presence of supernumerary ribs was associated with long gap oesophageal atresia though statistically insignificant, it needs to be studied further with multicenter collaboration

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Conflicts of interest

There are no conflicts of interest.

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