A STAGED APPROACH IN TRIPLE ATRESIA

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INTRODUCTION

Triple atresia a rare entity of VACTERL association comprises of esophageal atresia(EA), duodenal atresia(DA) and anorectal malformation(ARM)[1]. It is associated with high mortality[2]. It remains a challenge in surgical management whether to perform single staged or staged surgery for all three anomalies[1].

Any neonatal surgery is considered a stressor to neonates. Prolong surgery, hypothermia and metabolic changes may influence survival and surgical outcome[8].

Currently, there is no ideal approach for triple atresia as the incidence remains low. We are reporting a case of staged approach for triple atresia.

CASE SUMMARY

A term baby girl with a birth weight of 2.36kg was referred to us for excessive drooling of saliva. Antenatally mother had polyhydramnios. Clinically baby was not syndromic, not ventilated but required nasal prong oxygen, no murmur heard, abdominal examination was unremarkable, normal female genitalia but absent anal opening. We were unable to insert ryles tube.

X Ray revealed coiling of nasogastric tube at T2 level and the appearance of double bubble sign (figure 1) confirming the diagnosis of triple atresia.

Figure 1: chest & radiograph showing coiling of ryles tube at T2 and presence of double bubble sign

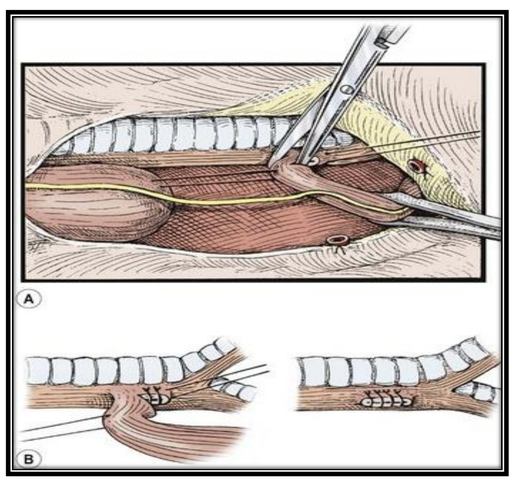

Figure 2 : Tracheoesophageal fistula ligation

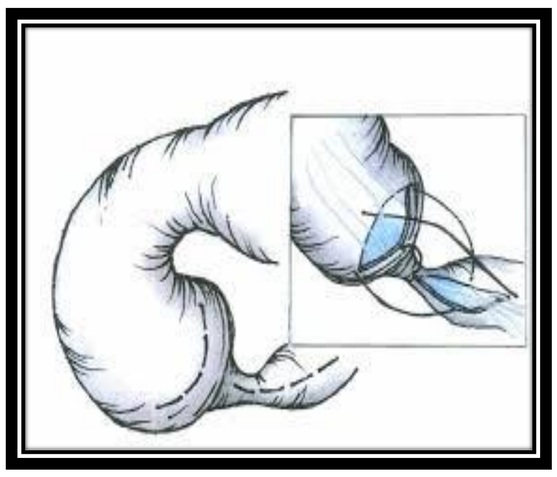

Figure 3: KIMURA duodenoduodenostomy

- Right thoracotomy was performed at day 3 of life. Through extra pleural space, the distal fistula was identified and ligated.
- Esophageal anastomosis was done without tension.
- Post operatively she was ventilated on low setting and hemodynamically supported with single inotrope.
- Day 7 of life she underwent laparotomy.
- Type 3 DA noted and Kimura duodenoduodenostomy was performed together with left transverse colostomy for her ARM which was placed at different incision.
- Postoperatively patient was on conventional ventilation before extubated after day 4 post operative.

Feeding was initiated after 7 days until achieved full feeding. Patient remained well under room air until she was discharged with omeprazole after 5 weeks post operative.

Currently she is awaiting definitive surgery for her ARM.

DISCUSSION

Triple Atresia(TA) poses a great dilemma for pediatric surgeon in diagnosing and tackling the complexity in planning the surgery. There are divided opinions whether to perform all anomalies in a single surgery or in stages. Even for staged surgery, the choice of either repairing EA first or prioritize DA or colostomy.

Staged surgery was the choice in most reported cases [1,5]. Raef et al stated better survival rate in staged surgery compared to those managed in single surgery.

As for now there is no clear guideline available in managing TA as the anomaly is extremely rare[1,3].

Neonates are susceptible to any stressors, any newborn subjected to surgery increases the risk of hypothermia, dehydration, hypotension, excessive metabolic changes, all of which interferes with a neonates normal homeostatis[7]. These are exaggerated by prolong surgery which is associated with post operative mortality[8].

It is well established in Lisanne et al's paper associating delayed neurodevelopmental in neonates that undergone major surgery[9].

Moreover staged surgery allows improvement in neonates cardiovascular and pulmonary function [3,5,6,7]. It could also allow neonatologist to optimize ventilator setting, inotropic support, fluid requirement and antibiotic regime. In Singh et al's study, staged surgery improved survival rate for patients associated with higher mortality such as prematurity, low birth weight, those in sepsis and cardiac disease.

Surgeons may also recover from mental and physical exhaustion before embarking to subsequent surgery.

We opted to perform EA with distal TOF repair first as there is risk of gastric distension and perforation which is associated with higher mortality in ventilated TOF[10]. In which, Raef and Spitz et al both found that performing TOF as first surgery produced better outcome [1,4].

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CONCLUSION

Triple Atresia is a rare condition and surgical approach remains challenging. In our point of view, staged operation is known to be the optimal approach for such cases.