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Letter to the Editor

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Triple atresia: Staged repair

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DEAR SIR

Triple atresia is a rare condition consisting of esophageal atresia, duodenal atresia, and anorectal malformation. It brings dilemmas for surgeons in diagnosing the condition itself and in planning for surgical management, whether to perform surgery in a single stage or in stages for all three anomalies. In staged surgery, there are options of either repairing esophageal atresia first or prioritizing duodenal atresia or colostomy. However, the surgery itself predisposes these neonates to risks of prolonged surgery, hypothermia, and metabolic changes that may affect the prognosis.

A term female neonate with a birth weight of 2.36 kg was referred to us for excessive drooling of saliva. Clinically the baby had no dysmorphism with unremarkable abdominal findings, and normal female genitalia but absent anal opening. Resistance was noted during Ryle's tube insertion. Infantogram revealed coiling of Ryle's tube at T2-3 vertebra level, prominent gastric bubble, and duodenal cap with no distal bowel gas (Figure 1). These findings made a provisional diagnosis of Type C esophageal atresia with distal tracheoesophageal fistula, duodenal atresia, and high-type anorectal malformation. Staged surgery began with right thoracotomy on day 3 of life through 4th intercostal space, with an extrapleural approach the fistula was ligated with prolene 5-0, and primary esophageal anastomosis completed using PDS 6-0. Postoperatively, the baby was shifted to NICU, ventilated on low settings, and hemodynamically supported with single inotropes.

On day 7 of life, she underwent laparotomy via a right upper transverse incision. A Type 3 duodenal atresia was identified and Kimura duodeno-duodenostomy was performed. A left transverse colostomy for anorectal malformation was also made. The baby was in NICU on conventional ventilation till day 11 of life. Ryle's tube feeding was initiated on day 7 postoperative. There was no episode of feeding intolerance. Feeding was gradually stepped up and was discharged with omeprazole after 5 weeks postoperative. Corrective surgery for anorectal malformation was performed at 1 year of age via the posterior sagittal route. There was a rectovaginal fistula noted. The fistula was detached and transfixed. Rectum was mobilized and anorectoplasty was done.

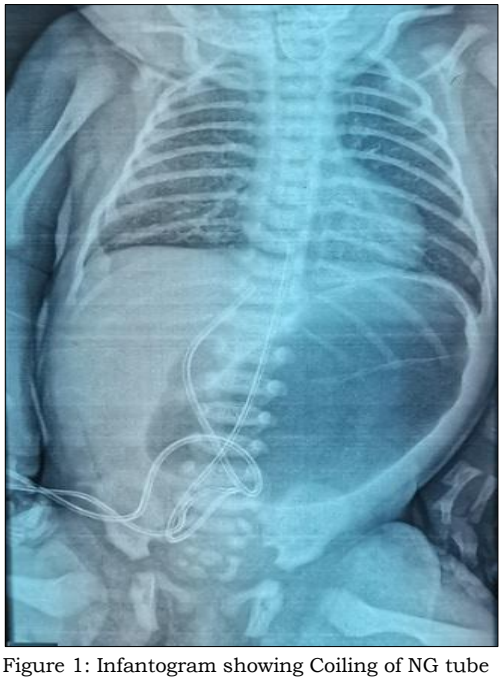

Figure 1: Infantogram showing Coiling of NG tube in the upper thorax and dilated stomach and duodenal cap with no distal gas.

Table 1 depicts a literature review of triple atresia published cases. It shows that seven cases had undergone staged repair of their respective anomalies in which 4 of them underwent laparotomy for bowel pathology first then followed by thoracotomy, ligation of fistula, and esophageal anastomosis. In the other 3 cases from the staged group, thoracotomy was performed prior to laparotomy. Meanwhile, there were 3 cases in the staged group that had concomitant congenital heart disease and in 2 cases, babies'

weights were under 2.0 kg. A single staged surgery was performed in 4 cases of which 3 of them, thoracotomy was performed first and followed by laparotomy. Unfortunately, 2 of them died within one week postoperatively.

Table 1: Summary of triple atresia literature review

Ref/ Year	Types of Atresia	Age / Weight/ Gender	Associated Anomalies	Sequence of Neonatal Surgery (Single/ Staged)	Outcome
Kawana 1989	1) OA TEF 2) ARM 3) DA	Day 1 old/ 2.6kg/ Male	Right radial aplasia	SINGLE STAGED: TEF ligation > primary repair OA > sigmoid colostomy > Duodeno-duodenostomy	Survived neonatal period
Harjai 2000	1) OA TEF 2) ARM 3) DA	Day 1 old/ 2.4kg/ Male	N/A	SINGLE STAGED: TEF ligation > primary repair OA > Duodeno-duodenostomy > gastrostomy > sigmoid colostomy	Died at day 8, had anastomotic leak
Panda 2015	1) OA TEF 2) ARM 3) DA	Day 2 old/ NA/ Male	N/A	STAGED: 1. Sigmoid loop colostomy > Duodeno-duodenostomy 2. TEF ligation > primary repair OA (after 72H)	Survived neonatal period
	1) OA TEF (Long gap) 2) ARM (Pouch colon) 3) DA	Day 4 old/ NA/ Female	- Atrial Septal Defect - Ventricular Septal Defect	STAGED: 1. Excision of Type IV pouch colon + ligation of colovesical fistula + end colostomy 2. TEF ligation + cervical esophagostomy (after 72H) 3. Duodeno-duodenostomy (after 96H)	Survived neonatal period
	1) OA TEF 2) ARM 3) DA	Day 3 old/ NA/ Male		STAGED: 1. Repair of posterior stomach wall perforation + gastrostomy + sigmoid colostomy 2. TEF ligation > primary repair OA (after 48H) 3. Duodeno-duodenostomy (after 9 days)	Survived neonatal period.
	1) Pure OA 2) ARM (Pouch Colon) 3) DA with malrotation	Day 2 old/ NA/ Female	N/A	SINGLE STAGED: Sigmoid colostomy > ligation of colovesical fistula > cervical esophagostomy > gastrostomy > Ladd's procedure and Duodeno-duodenostomy	Survived neonatal period
	1) OA TEF 2) ARM 3) DA	Day 5 old/ NA/ Male	Cardiac: - Atrial Septal Defect - Ventricular Septal Defect, PDA Radial Aplasia	STAGED: 1. Sigmoid loop colostomy 2. TEF ligation > primary repair OA (after 96H) 3. Duodeno-duodenostomy + gastrostomy (after 72H)	Survived neonatal period
Khanna 2017	1) OA TEF 2) Pyloric web 3) ARM	Day 2 old/ NA/ Male	N/A	SINGLE STAGED: TEF ligation > primary repair OA > laparotomy and pyloroplasty > sigmoid colostomy.	Died POD 6 - sepsis, pneumonia
Vinod 2018	1) Pure OA 2) ARM 3) DA	Day 1 old/ 1.6kg/ Female	N/A	STAGED: 1. Cervical Esophagostomy (day 2 of life) 2. Duodeno-duodenostomy (day5 of life)	Died POD 4 - septic shock
Raef 2021	1) OA TEF 2) ARM 3) DA	Day 1 old/ 1.7kg/ Male	Cardiac: - Atrial Septal Defect - PDA	STAGED: 1. TEF ligation > primary repair OA (day2 of life) 2. Duodeno-duodenostomy and sigmoid colostomy (day7 of life)	Survived neonatal period
	1) OA TEF 2) ARM 3) DA	Day 1 old/ 2.4kg/ Female	N/A	STAGED: 1. TEF ligation > primary repair OA (day2 of life) 2. Duodeno-duodenostomy (day4 of life)	Survived neonatal period
Our case	1) OA TEF 2) ARM 3) DA	Day 1 old/ 2.36kg/ Female	No	STAGED: 1. TEF ligation > primary repair OA (day3 of life) 2. Duodeno-duodenostomy and left transverse colostomy (day 7 of life)	Survived neonatal period

This experience convinces us that the decision to perform TEF repair first and followed by laparotomy and stoma is safer and offers better survival. Raef et al favor staged surgery over single-staged surgery as it is found to have a better survival rate.[1] As outlined in table 1, only one mortality was documented among the staged group. However, as of now, there is no clear guideline available for managing triple atresia as the anomaly remains extremely rare.[1-3] As to the sequence of staged surgery, we opted to perform TEF repair first as there is a risk of gastric distension due to air venting down from the trachea into the stomach through distal fistula which can also cause gastric perforation.[4] Both Raef and Spitz et al suggest that performing TEF repair as the first surgery yields a better outcome.[1,5].

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