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The Thoracic Mishap of Tef with Cdh: A Case Report

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1. Abstract

1.1. Background:

Coexistence of tracheo-osophageal fistula with congenital diaphragmatic hernia is very rare. There are only 36 reported cases till date and no specific consensus established regarding management. But working on their combined pathophysiology meticulously; peri-operative,anaesthetic and surgical challenges in management of the duo can be tackled with available resources.

1.2. Case Presentation:

We present case of full term, small for gestational age neonate with birth weight of 2.100 kg having type C Tracheo-osophageal fistula and left Bochdalek type congenital diaphragmatic hernia with persistent pulmonary hypertension. Baby was managed in neonatal intensive care unit with intravenous sildenafil and high frequency oscillatory ventilator. Baby was operated on day 2 for repair of diaphragmatic hernia with left tube thoracostomy and gastro-esophageal junction ligation, feeding gastrostomy, prosthetic patch abdominoplasty and left cervical esophagostomy. On post Opday 2, baby's condition deteriorated and could not be revived.

1.3. Conclusion:

Though there are less reported cases and lacking specific protocols for management of 'tracheo-osophageal fistula with congenital diaphragmatic hernia', treatment can be individualized considering patient's condition and available medical services.

2. Introduction

Combination of Tracheo-Osophageal Fistula (TEF) with Congenital Diaphragmatic Hernia (CDH) was considered almost non-existent once. But till date 36 cases are reported in literature. We present the India's first reported case of type C TEF with left Bochdalek type CDH; also discuss its challenges and various ways of management.

As both entities exhibit their own patho-physiologies, we have to plan medical and surgical management very meticulously taking all available facilities into consideration. Considering surgical management, we have to do either 'spaced' or 'at the same time' repair of both anomalies as per patient's condition.

This combination is challenging at every management step but not always deadly. With use of current advancement in neonatal medical facilities, this rare duo can be managed well.

3. Case Report

We present case of a neonate who was full term, caesarean delivery (in view of previous caesarean delivery), and small for gestational age, with birth weight of 2.100 kg. The baby was antenatally diagnosed with polyhydramnios with Amniotic Fluid Index (AFI) of 20 cm, thoracic hemivertebrae and left CDH at 24 weeks of gestation. The baby was borne to 27 years old lady who was gravida 3, para 2 with 1 abortion (at 2 months of gestation) on her obstetric history.

Baby cried immediately after birth. In view of tachypnea, baby was intubated at 2nd minute after birth. When Nasogastric (NG) tube insertion was attempted, we couldn't pass it beyond mark of 10 cm. The chest and abdomen x ray (Figure1) revealed NG tube reaching up to T4 vertebral level (suggestive of TEF!!!), heart shadow pushed to right side and bowel loops in left thoracic region. So, we found out that we are dealing with 'TEF with CDH'!!!

In NICU, baby required intravenous sildenafil and High Frequency Oscillatory Ventilator (HFOV) in view of Persistent Pulmonary Hypertension (PPHN). Baby was operated on day 2 of life. Intra-operative findings were type C TEF with left Bochdalek type CDH. As patient's vitals deteriorated intra-operatively, plan of thoracotomy with fistula ligation was deferred at that time and after

left CDH repair with left tube thoracostomy (Figure 2); gastro-esophageal junction ligation, feeding gastrostomy, abdominoplasty with silo (to avoid intra-abdominal hypertension) and left cervical esophagostomy were performed. On post-operative Day 2, baby's PPHN worsened, developed refractory cardiogenic shock and despite maximal resuscitation efforts, baby could not be revived.

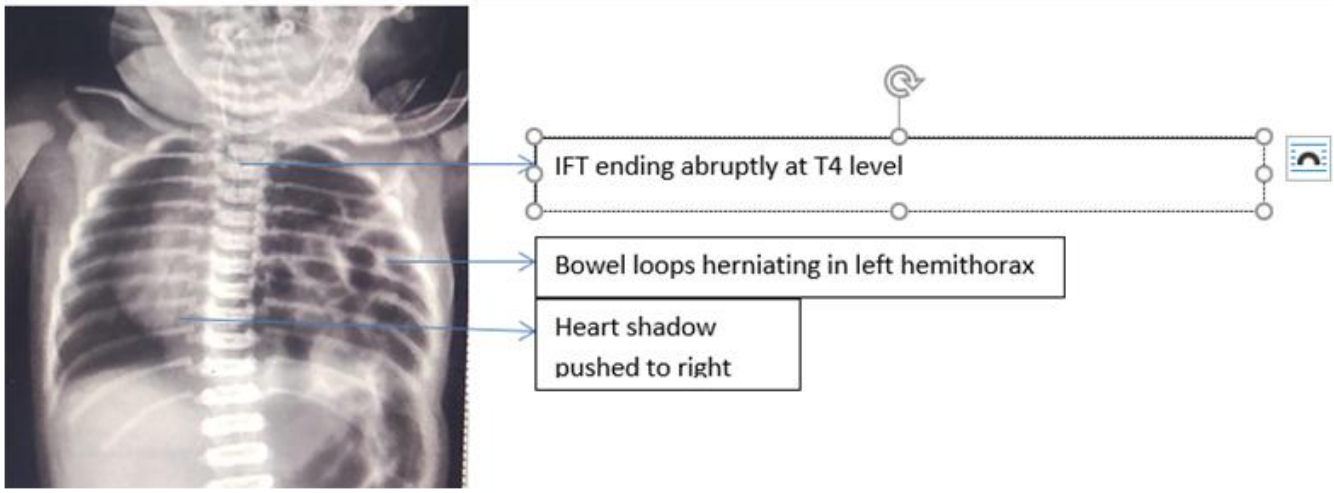

Figure 1

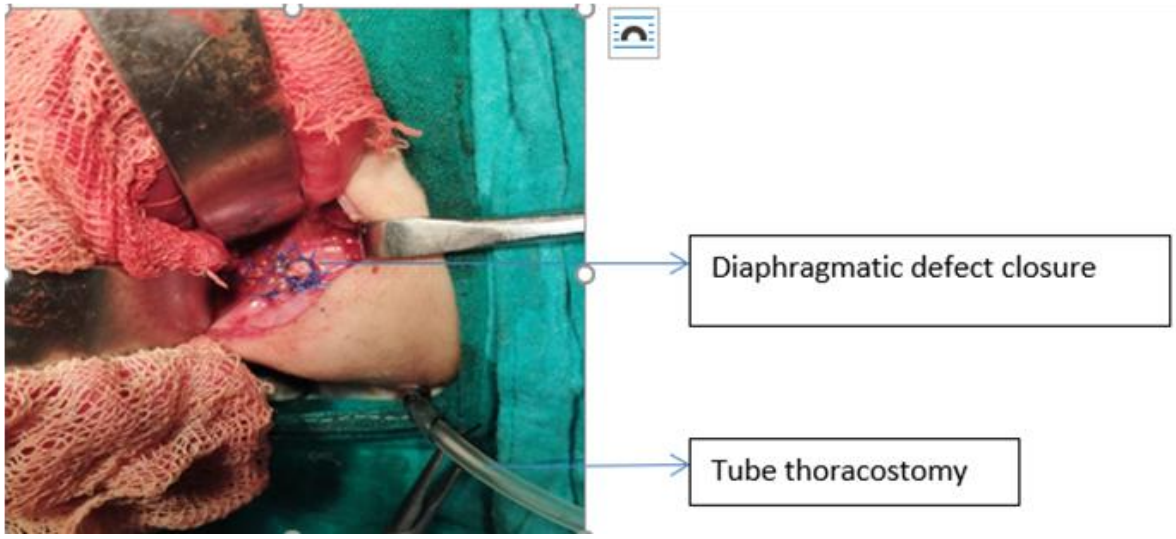

Figure 2

4. Discussion

Incidence of co-existence of both "TEF and CDH" is less than 1 in 4500 live births[1]. TEF occurs in less than 10% CDH [2] while focal muscular deficiency of diaphragm leading to eventration occurs in 10-50% of TEF but not as typical Bochdalek type hernia[2].

'TEF with CDH' manifests with combination of pathophysiology of both. CDH contributes for PPHN, ipsilateral and sometimes contralateral lung hypoplasia, poor pulmonary compliance, intestinal rotational anomalies while TEF adds by risk of rapid and uncontrolled gastro-intestinal dilation with significant respiratory volume loss in relatively low pressured gastrointestinal tract [3], risk of aspiration and chemical pneumonitis.

Management of the duo is challenging at every step.

The first tricky job is "how to tackle air leak into gastrointestinal track (GIT)???" and evidence based solutions to this question are 1) Passing NG tube through fistula purposefully and decompressing GIT [2], 2) Bronchoscopy guided fogarty balloon occlusion of fistula (but with due risk of tracheal occlusion by balloon slippage) [2], 3) selective endo-bronchial intubation of right main bronchus (bronchus of relatively less hypoplastic lung) so that right lung is well ventilated, hypoplastic left lung is not ventilated and side wall of ET tube occludes fistula [2], 4) Intubating gut by passing NG tube through Endotracheal (ET) tube through T connector [3].

The next challenge is" whether to do all repairs at once or go for spaced repairs?"

Thakral et al [4] stated that outcome of operating both anomalies early and simultaneously gives outcome as good as when dealing with either single anomaly. While Bagci et al [5] mentioned that considering poor pulmonary reserve; Esophageal Atresia (EA)

can be approached at later date with due precautions to prevent aspiration pneumonia.

Ahmed et al [6] emphasized on early gastrostomy but Fann et al [7] explained “waterseal gastrostomy”.

The next challenge is how to address ‘volume loss’ in case if ‘thoracotomy with fistula ligation’ isn’t possible.

Leininger et al [8] elaborated on simple ligation of distal esophagus (done to prevent volume loss) complicating into stricture. While to avoid “stricture complication” Fagelman et al [9] proposed temporary banding of gastroesophageal junction with elastic silicone rubber and its future removal without surgery.

The next issue is ‘to achieve abdominal closure (in case of abdomino-visceral disproportion) without risking patient for intra-abdominal hypertension’.

For that Zahn et al [10] proposed silastic bag closure of abdomen.

In our case, when the patient’s condition was favorable to proceed with surgery, we had planned for left CDH repair via left subcostal incision with prosthetic patch abdominoplasty, right thoracotomy to ligate the fistula and primary esophageal repair. But as the patient’s vitals worsen intra-operatively, plan of thoracotomy was abandoned and instead gastro-oesophageal junction ligation with diversion procedures of TEF was performed. Due to refractory cardiogenic shock, we lost the patient on second post-operative day.

5. Conclusion

There is no specific consensus established for management of ‘TEF with CDH’ probably due to its rarity and less reported cases. But on basis of available literature; treatment can be individualized with panoramic view of entire scenario- “severity of PPHN, presence or absence of fistula in case of TEF, patient’s general and systemic conditions and availability of medical services”.

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