

Case Report

## Right Phrenic Nerve Injury: A Rare but Possible Complication of Oesophageal Atresia and Tracheoesophageal Fistula Repair

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**Abstract** — Oesophageal atresia (OA) and Tracheoesophageal fistulas (TOF) are congenital anomalies commonly present in between 1 and 4 per 10 000 births, including stillbirths and termination of pregnancies. Currently, patients with these anomalies are managed surgically, with common complications including oesophageal formation, anastomotic leakage and recurrence of TOF. This case report highlights the possibility of right phrenic nerve injury as a complication of OA and TOF repair. We report a baby boy born at term with worsening respiratory distress at day 15 of life, two weeks after undergoing an operation to repair OA and TOF. Since the surgery on day 2 of life, he could not be weaned off oxygen support successfully and had two episodes of worsening respiratory distress that was treated as pneumonia. Serial Chest X-Rays (CXR) showed a persistent elevation of the right hemidiaphragm. Ultrasound (USG) thorax supported the findings of right diaphragmatic paralysis by revealing minimal right diaphragm excursion with the paradoxical movement of the right diaphragm. He was initially managed non-operatively with non-invasive ventilation (NIV)/continuous positive airway pressure (CPAP) while monitoring for spontaneous recovery of the right diaphragm, which unfortunately did not occur. He was on NIV/CPAP for a total of 25 days and was tachypnoeic with mild subcostal recessions (SCR) and reduced breath sounds on the right throughout. Six weeks after the initial OA/TOF repair surgery, he was posted for Right Diaphragmatic Plication. Afterwards, he recovered well and was weaned off oxygen support within the next seven days and subsequently discharged.

**Keywords** — Phrenic Nerve Palsy, Oesophageal Atresia, Tracheoesophageal fistula, non-operative management, Thermal injury, Diaphragmatic Plication

### I. INTRODUCTION

Oesophageal atresia (OA) and Tracheoesophageal fistulas (TOF) are congenital anomalies commonly present together, ranging between 1 and 4 per 10 000 births, including stillbirths and termination of pregnancies [1]. Currently, patients with these anomalies are managed surgically as the trachea and oesophagus are separated via ligation of the interconnecting fistula. In cases of OA, both oesophageal

segments are anastomosed. The most common complications resulting from the repair of TOF and OA that have been reported include oesophageal stricture formation, anastomotic leakage, surgical site infection, postoperative shock or sepsis and recurrence of TOF [2]. In addition, this case report wishes to highlight the possibility of right phrenic nerve injury as a complication of OA and TOF repair.

## II. CASE REPORT

Our patient, a baby boy, was born at term (39W 2D) via vacuum-assisted delivery following an abnormal CTG with a birth weight of 2.35kg and a good APGAR score. He was initially admitted due to abnormal cord blood. However, when examined at 2 hours of life, he was tachypnoeic with a respiratory rate (RR) of 50/min, intermittent nasal flaring and mild subcostal recession (SCR), but his oxygen saturation (SPO<sub>2</sub>) under room air (RA) was 100%. He was put on nasal prong oxygen (NPO<sub>2</sub>) at 1L/min and covered for presumed sepsis in view of his respiratory distress. Significant pooling of oral secretion was seen during the attempted insertion of an orogastric tube, which was difficult, with significant resistance being felt on insertion. A coiled Ryles tube was seen on a chest X-Ray (CXR) that was done afterwards, as shown in Figure 1.



Figure 1 AP CXR taken at 5 hours of life showing coiling of Ryles tube (white arrow)

He was then started on continuous oral suction using a Replogle tube and was subsequently posted for Right Thoracotomy with TOF ligation by the Paediatric Surgery team. During the procedure, an incision was marked 1cm below the scapula extending from the mid-axillary line to the paravertebral region posteriorly. The thorax was then entered at the 4th intercostal space by splitting the intercostal muscles. A Finochietto (rib spreader) was used to spread the ribs gently. A retractor was used to retract the lung and expose the azygos vein, which was then ligated and divided using Vicryl 4/0. Intraoperatively, the surgeons noted that our patient had Oesophageal atresia (OA) with distal TOF Gross type C [3]. The fistula was then ligated, and the upper pouch was identified and mobilised extensively to bridge the gap for anastomosis. PDS 5/0 was used to secure both upper and lower pouches. The surgery was uneventful, with minimal blood loss. One hour postoperatively, upon arrival at the NICU, the right chest wall was noted to be higher than the left side. Auscultation revealed reduced air entry on the right side, and his SPO<sub>2</sub> was at 80%. Arterial blood gas revealed mild metabolic acidosis, and he was subsequently kept on

ventilatory support. A CXR done on postoperative day (POD) 3 noted right upper lobe (RUL) collapse, as seen in Figure 2.



Figure 2 AP CXR taken on POD3 showing RUL collapse.

Over the next few days, the child improved clinically but was unable to be weaned off from O<sub>2</sub> support completely with several episodes of worsening respiratory distress, which was managed with antibiotics and escalating his oxygen support from NPO<sub>2</sub> to high-flow nasal cannula (HFNC) at 5-6L/min. Two CXRs done within that period noted bilateral pleural effusion with worsening right lung consolidation, slowly improving by POD 15. However, both films also revealed a persistently elevated right hemidiaphragm seen as three ribs higher than normal, as shown in Figure 3.

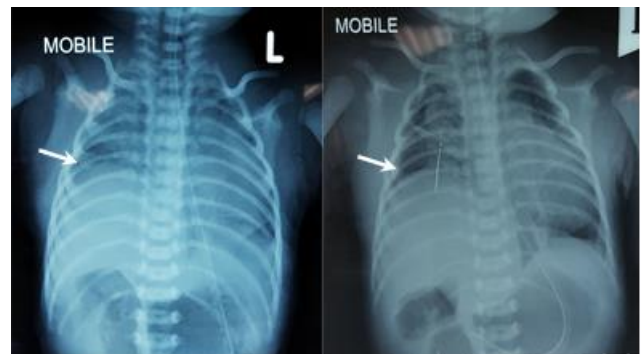


Figure 3 CXRs taken on POD 12 and 15 which showed a persistently elevated right hemidiaphragm (white arrows). Additionally, a bilateral pleural effusion with right lung consolidation is seen on POD 12 film (shown on the left) that is improved by POD 15 (shown on the right).

Ultrasonography of the thorax done one day later showed minimal right diaphragm excursion and paradoxical movement of the right diaphragm, which were suggestive of right diaphragmatic paralysis. Following this, for a total of 25 days (POD19 to POD44), our patient could not be weaned off DuoPAP/CPAP and was seen to have baseline tachypnoea and SCR all throughout. Apart from that, CXR on POD 31 still

noted a persistently elevated right hemidiaphragm and some consolidation over the right lung, similar to what is seen in Figure 3, as shown above. Therefore, the Paediatric Surgery team decided to post our patient for right diaphragmatic plication if diaphragmatic eventration was still seen at 6 weeks post-op. Unfortunately, the CXR done 6 weeks post-op revealed a persistently elevated right hemidiaphragm yet again, as shown in Figure 4, and the patient was posted for the abovementioned surgery.



Figure 4 AP CXR taken 6 weeks after the initial TOF repair surgery still showing a persistently elevated right hemidiaphragm (white arrow)

Fortunately, the procedure went on smoothly, and post-plication, the right diaphragm showed good contraction and relaxation. Furthermore, another CXR done soon after the procedure confirmed the position of both the right and left hemidiaphragm at the appropriate anatomical level (T8-T9), as seen in Figure 5.



Figure 5 AP CXR taken shortly after Right Diaphragmatic Plication showing both hemidiaphragms at the correct anatomical position of T8-T9.

On examination postoperatively, the child was not tachypnoeic with minimal SCR, and the lungs were clear with equal air entry bilaterally. His recovery postoperatively was uneventful as he was extubated to NPO2 1L/min the very next

day and successfully weaned off oxygen support 7 days later. He was discharged shortly afterwards and given a follow-up appointment at the Paediatric Surgical Clinic in one month.

### III. DISCUSSION

The common postoperative complications of Oesophageal atresia (OA) and Tracheoesophageal fistulas (TOF) repair include anastomotic strictures, gastro-oesophageal reflux, anastomotic leak, recurrent TOF, tracheomalacia and respiratory infections [4]. However, cases of iatrogenic phrenic nerve injury with resultant diaphragmatic paralysis have been reported in the past, albeit very rarely [5-7]. After entering the thorax via the superior thoracic aperture, the right phrenic nerve descends anteriorly to the right lung root and along the pericardium of the heart's right atrium before piercing the diaphragm to innervate its inferior surface. It is a frequently cited complication of paediatric cardiac surgery for this reason [8-10]. In this case, direct injury to the phrenic nerve was unlikely due to the anatomical location of the incision, which is posterolateral, extending from the mid-axillary line to the paravertebral region. Instead, it is postulated that this rare complication occurred because of indirect injury to the phrenic nerve, contributed by multiple factors. Firstly, like many patients who undergo this surgery, our patient was small (birth weight 2.35kg), allowing for minimal intervening tissue, thereby increasing the possibility of lateral thermal damage and retraction injury at relatively distant sites [5]. Secondly, heat spread to adjacent tissues from the use of electrocautery might have resulted in the indirect thermal injury of the phrenic nerve depending on the application time, type of device used, or the power settings applied. Thirdly, the particularly high location of the upper pouch of the oesophagus in the mediastinum might have caused traction exerted on the phrenic nerves or its cervical nerve roots during its mobilisation, which was significant and extensive [5, 6].

In paediatric patients, diaphragmatic paralysis tends to be symptomatic and frequently manifests as dyspnoea, tachypnoea, failed extubation or recurrent reintubation, prolonged ventilation, pneumonia, and sepsis, as seen in our patient [11]. Therefore, affected patients with diaphragmatic paralysis may be given positive-pressure ventilation for longer, in the hopes of functional recovery and avoiding additional surgery. However, the recovery of a phrenic nerve palsy is unpredictable, with studies quoting recovery periods ranging from days to months [9, 12, 13]. Moreover, prolonged intensive care unit (ICU) admission can put vulnerable patients at risk of additional morbidity and mortality, thus favouring early surgical intervention [14]. Hence, many authors recommend early diaphragmatic plication (less than 14 days from the index surgery) as it has been shown to result in earlier extubation (within 2 to 3 days on average), shorter ICU and hospital stays, lower morbidity, and thus improved outcomes [14-17].

However, in our case, the likelihood of an indirect injury is very high, as mentioned earlier. From the few reports published regarding phrenic nerve palsy following OA/TOF repair, all recovered following several weeks of non-operative management, primarily via CPAP. For instance, Henderson et al. reported a case of right hemidiaphragm paralysis following

OA/TOF repair, which required the affected baby to be on ventilatory support for 7 weeks (34 days of which on CPAP) before resolving spontaneously [5]. A similar case reported by Man et al. required 5 weeks of ventilatory support in the form of IPPV/CPAP, after which spontaneous recovery was noted [6]. Another similar case reported by Haller et al. also noted spontaneous recovery following several weeks of ventilatory support via CPAP [7].

Additionally, diaphragmatic plication is not entirely risk-free, with concerns about the function of the plicated diaphragm in the long run. While studies have shown that plication does not prevent the return of diaphragmatic motion in the years following the procedure [18], smaller studies that have followed children who underwent diaphragmatic plication have found that this procedure ultimately results in normal or close to normal diaphragmatic function after 1 to 7 years with some children unfortunately never regaining normal function within that time period [9, 17, 19]. However, Lemmer et al. noted that while children with plicated diaphragms may have lower lung function values, they usually have a normal exercise capacity [20].

Considering all that, we believe that young patients (newborns and infants) who are respiratory support dependent, and especially those with comorbidities involving respiratory, central nervous and cardiovascular systems will more likely need to undergo diaphragmatic plication. Note that an early plication should be considered in those patients due to their reduced ability to tolerate diaphragmatic paralysis [21]. For our patient, plication was carried out 6 weeks after the index surgery as the management team was hoping for the spontaneous recovery of his diaphragmatic paralysis due to the indirect nature of the injury, which ended up not improving throughout the observation period. In hindsight, it might have been better for our patient to have had his paralysed right diaphragm plicated well before complications of prolonged ventilatory assistance set in. It should be noted that our patient underwent a traditional open transthoracic plication as opposed to other minimally invasive techniques, which increases the risk of long-term complications such as scoliosis [22]. These children should then be followed up every six months or yearly as needed to monitor their lung function and watch out for complications, as mentioned earlier.

#### IV. CONCLUSIONS

Phrenic nerve injury should be considered as one of the complications of Oesophageal atresia (OA) and Tracheoesophageal fistula (TOF) repair, especially when the patient has difficulty being weaned off from ventilatory support post-op. It is important to note that paralysis of the hemidiaphragm, in this case, was most likely caused by indirect injury to the phrenic nerve. Therefore, similar patients should be given a trial of non-operative management via ventilatory support while monitoring for spontaneous recovery. If they are unable to be weaned off ventilatory support, early diaphragmatic plication should be considered in view of the increased morbidity and mortality rates of prolonged hospital stays.

#### CONSENT TO PARTICIPATE

Written informed consent was obtained from the patient for the anonymized information to be published in this article.

#### CONFLICT OF INTERESTS

The authors declared no conflict of interest and did not receive any funding for this article.

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The authors' contributions were as follows- SS and VNS reviewed current literature, drafted, and wrote the manuscript. AK and RMS edited, reviewed, and approved the final version of the article. AK, MFS and QL provided input from a surgical perspective.

#### REFERENCES

- [1] Brosens, E., Ploeg, M., van Bever, Y., Koopmans, A.E., IJsselstijn, H., Rottier, R.J., Wijnen, R., Tibboel, D. and de Klein, A., 2014. Clinical and etiological heterogeneity in patients with tracheo-esophageal malformations and associated anomalies. *European journal of medical genetics*, 57(8), pp.440-452. doi.org/10.1016/j.ejmg.2014.05.009
- [2] Lal, D.R., Gadepalli, S.K., Downard, C.D., Ostlie, D.J., Minneci, P.C., Swedler, R.M., Chelius, T., Cassidy, L., Rapp, C.T., Deans, K.J. and Fallat, M.E., 2017. Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula. *Journal of pediatric surgery*, 52(8), pp.1245-1251. doi.org/10.1016/j.jpedsurg.2016.11.046
- [3] Gross, E. R. (1962). *The surgery of infancy and childhood*.
- [4] Lee, S., & Meeker, T. M. (2018). Basic knowledge of tracheoesophageal fistula and esophageal atresia. *Advances in Neonatal Care*, 18(1), 14-21. doi.org/10.1097/ANC.0000000000000464
- [5] Henderson, P. W., & Spigland, N. A. (2010). Right phrenic nerve injury as a complication of tracheoesophageal fistula repair. *Pediatric Critical Care Medicine*, 11(5), e52-e54. doi.org/10.1097/PCC.0b013e3181dde5c3
- [6] Man, D., Wheildon, M. H., & Eckstein, H. B. (1982). Paralysis of the right hemidiaphragm following primary anastomosis for oesophageal atresia and tracheo-oesophageal fistula. *Zeitschrift für Kinderchirurgie*, 37(09), 32-33. doi.org/10.1055/s-2008-1059812
- [7] Haller Jr, J. A., Pickard, L. R., Tepas, J. J., Rogers, M. C., Robotham, J. L., Shorter, N., & Shermeta, D. W. (1979). Management of diaphragmatic paralysis in infants with special emphasis on selection of patients for operative plication. *Journal of pediatric surgery*, 14(6), 779-785. doi.org/10.1016/S0022-3468(79)80264-X
- [8] Mok, Q., Ross-Russell, R., Mulvey, D., Green, M., & Shinebourne, E. A. (1991). Phrenic nerve injury in infants and children undergoing cardiac surgery. *Heart*, 65(5), 287-292. dx.doi.org/10.1136/hrt.65.5.287
- [9] Watanabe, T., Trusler, G. A., Williams, W. G., Edmonds, J. F., Coles, J. G., & Hosokawa, Y. (1987). Phrenic nerve paralysis after pediatric cardiac surgery: retrospective study of 125 cases. *The Journal of Thoracic and Cardiovascular Surgery*, 94(3), 383-388. doi.org/10.1016/S0022-5223(19)36251-8

- [10] Fraser III, C.D., Ravekes, W., Thibault, D., Scully, B., Chiswell, K., Giuliano, K., Hill, K.D., Jacobs, J.P., Jacobs, M.L., Kutty, S. and Vricella, L., 2021. Diaphragm paralysis after pediatric cardiac surgery: An STS Congenital Heart Surgery Database study. *The Annals of Thoracic Surgery*, 112(1), pp.139-146. doi.org/10.1016/j.athoracsur.2020.05.175
- [11] Parmar, D., Panchal, J., Parmar, N., Garg, P., Mishra, A., Surti, J., & Patel, K. (2021). Early diagnosis of diaphragm palsy after pediatric cardiac surgery and outcome after diaphragm plication—A single-center experience. *Annals of Pediatric Cardiology*, 14(2), 178. doi.org/10.4103/apc.APC\_171\_19
- [12] Mickell, J. J., Oh, K. S., Siewers, R. D., Galvis, A. G., Fricker, F. J., Mathews, R. A., & Bahnson, H. T. (1978). Clinical implications of postoperative unilateral phrenic nerve paralysis. *The Journal of Thoracic and Cardiovascular Surgery*, 76(3), 297-304. doi.org/10.1016/S0022-5223(19)39570-4
- [13] Iverson, L. I., Mittal, A., Dugan, D. J., & Samson, P. C. (1976). Injuries to the phrenic nerve resulting in diaphragmatic paralysis with special reference to stretch trauma. *The American Journal of Surgery*, 132(2), 263-269. doi.org/10.1016/0002-9610(76)90058-1
- [14] Floh, A. A., Zafurallah, I., MacDonald, C., Honjo, O., Fan, C. P. S., & Laussen, P. C. (2017). The advantage of early plication in children diagnosed with diaphragm paresis. *The Journal of Thoracic and Cardiovascular Surgery*, 154(5), 1715-1721. doi.org/10.1016/j.jtcvs.2017.05.109
- [15] Affatato, A., Villagra, F., De Leon, J.P., Gomez, R., Checa, S.L., Vellibre, D., Sanchez, P., Ji, D.B. and Brito, J.M., 1988. Phrenic nerve paralysis following pediatric cardiac surgery. Role of diaphragmatic plication. *The Journal of Cardiovascular Surgery*, 29(5), pp.606-609.
- [16] Hamilton, J. R. L., Tocewicz, K., Elliott, M. J., De Leval, M., & Stark, J. (1990). Paralyzed diaphragm after cardiac surgery in children: value of plication. *European journal of cardiothoracic surgery*, 4(9), 487-491. doi.org/10.1016/1010-7940(90)90171-U
- [17] Baker, C. J., Boulom, V., Reemtsen, B. L., Rollins, R. C., Starnes, V. A., & Wells, W. J. (2008). Hemidiaphragm plication after repair of congenital heart defects in children: quantitative return of diaphragm function over time. *The Journal of Thoracic and Cardiovascular Surgery*, 135(1), 56-61. doi.org/10.1016/j.jtcvs.2007.09.031
- [18] Schwartz, M. Z., & Filler, R. M. (1978). Plication of the diaphragm for symptomatic phrenic nerve paralysis. *Journal of pediatric surgery*, 13(3), 259-263. doi.org/10.1016/S0022-3468(78)80397-2
- [19] Stone, K. S., Brown, J. W., Canal, D. F., & King, H. (1987). Long-term fate of the diaphragm surgically plicated during infancy and early childhood. *The Annals of thoracic surgery*, 44(1), 62-65. doi.org/10.1016/S0003-4975(10)62359-4
- [20] Lemmer, J., Stiller, B., Heise, G., Alexi-Meskishvili, V., Hübler, M., Weng, Y., & Berger, F. (2007). Mid-term follow-up in patients with diaphragmatic plication after surgery for congenital heart disease. *Intensive care medicine*, 33(11), 1985-1992. doi.org/10.1007/s00134-007-0717-9
- [21] Goldberg, L., Krauthammer, A., Ashkenazi, M., Soudack, M., Tokatly Latzer, I., Vardi, A., & Paret, G. (2020). Predictors for plication performance following diaphragmatic paralysis in children. *Pediatric Pulmonology*, 55(2), 449-454. doi.org/10.1002/ppul.24539
- [22] Snyder, C. W., Walford, N. E., Danielson, P. D., & Chandler, N. M. (2014). A simple thoroscopic plication technique for diaphragmatic eventration in neonates and infants: technical details and initial results. *Pediatric surgery international*, 30(10), 1013-1016. doi.org/10.1007/s00383-014-3580-0