Sir Salimullah Medical College Journal

Sir Salimullah Med Coll J 2022; 30: 96-99

Case Report

Congenital Oesophageal Stenosis Diagnosed at 15 Days of Age: A Rare Case Report

Shabnam Shahidullah[1], Naima Sultana[2], Aditi Sarkar[3], Vikarun-Nesa[4] Sunirmal Roy[5], Nurun Nahar Begum[6]

Article information
Received: 10-10-2021
Accepted: 05-12-2021

Cite this article:
Shahidullah S, Sultana N, Sarkar A, Nesa V, Roy S, Begum NN. Congenital Oesophageal Stenosis Diagnosed at 15 Days of Age: A Rare Case Report. Sir Salimullah Med Coll J 2022; 30: 96-99

Key words:
Congenital esophageal stenosis (CES), Tracheoesophageal fistula (TEF), Neonatal age, Excessive Secretion, Neonatal Intensive Care Unit (NICU)

Abstract:

Congenital oesophageal stenosis is a rare entity with a frequently delayed diagnosis. Patients are often treated according to diagnosis of GERD until intolerance to semisolid diet appears and CES is suspected. It usually presents in infancy and childhood. We are reporting a case of congenital 0esophageal stenosis in fifteen days old male child. It is very rare to diagnose congenital oesophageal stenosis at this age.

Introduction :

Congenital esophageal stenosis (CES) is a rare anomaly of the esophagus with clinical significane and incidence is 1:25000 to 50000 live births. The defects results from incomplete separation of primitive foregut at 25th day of fetal life.[1] Diagnosis is usually delayed who presents in infancy and childhood. Frequent vomiting or regurgitation are key symptoms to suspect an abnormality of the esophagus. Some signs that should guide the clinician to an early diagnosis are difficulty in food ingestion and dysphagia, especially when the diet changes from breast feeding or commercialized milk to solid foods, along with repeated signs of choking during food ingestion.[2] But this patient presented with excessive secretion and feeding difficulty. We are reporting such a rare case. In our knowledge this is an earliest presentation of CES.

1. Asst. Registrar, Department of Paediatrics, Sir Salimullah Medical College, Mitford Hospital, Dhaka.
2. Indoor Medical Officer, Department of Paediatrics, Sir Salimullah Medical College, Mitford Hospital, Dhaka.
3. Registrar, Department of Neonatology, Sir Salimullah Medical College, Mitford Hospital, Dhaka.
4. Asst. Registrar, Department of Neonatology, Sir Salimullah Medical College, Mitford Hospital, Dhaka.
5. Professor, Department of Neonatology, Sir Salimullah Medical College, Mitford Hospital, Dhaka.
6. Professor, Department of Neonatology, Sir Salimullah Medical College, Mitford Hospital, Dhaka.

Correspondence : Dr Shabnam Shahidullah, Asst Registrar, Department of Paediatrics, Sir Salimullah Medical College, Mitford Hospital, Dhaka. sshabnam127@gmail.com

Case Report :

A 3 days old male preterm (35 weeks of gestation) baby delivered by LUCS due to placenta previa presented at 72 hours of age to NICU of Mitford Hospital with excessive secretion from mouth and unable to feed since birth. Baby cried immediately after birth. Physical Examination revealed weight 2400 gram, vitals are within normal limit except tachypnoea (respiratory rate -66 breaths/min). Air entry was poor on both lung field. Reflex activity was good and there was no apparent congenital anomaly. Our initial diagnosis was Preterm (35weeks) low birth weight (2400gm) early onset neonatal sepsis (EONS) with suspected Trache-oesophageal Fistula (TEF).

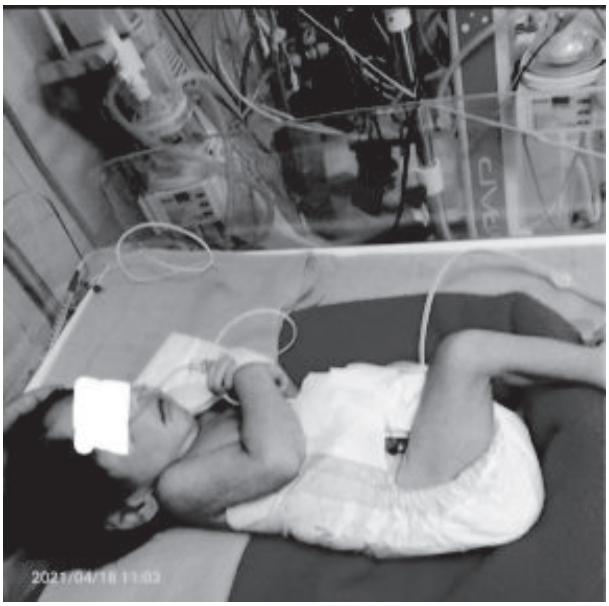

Fig.-1 : *A 3 days old new born having oesophageal atresia*

As a part of physical examination nasogastric (NG) tube was introduced. During procedure physician found some difficulties NG tube was passed in some extent. Baby was initially managed with nothing per oral, broad spectrum antibiotics, high flow oxygen supplementation, and other supportive therapy.

At that time some investigations were done and found and septic screening negative and serum Electrolytes were within normal limit. X-ray chest with NG tube in situ reveals no coiling of NG tube.

After three days, secretion from mouth and oxygen demand were reduced than before. But two days later patient again developed excessive secretion per mouth and respiratory distress in the form of tachypnoea .Treatment was given accordingly with fourth generation antibiotics and other supportive therapy. At that time diagnostic dilemma had been arisen. Then Upper GIT contrast X-ray was done that revealed – contrast medium has outlined the upper part of the oesophagus up to the level of D-4 with rounded lower part and proximal dilatation and diagnosed as oesophageal stenosis. The baby was then referred to Paediatric surgery Department for surgical purpose. But during routine check-up, he became covid positive and was treated accordingly.

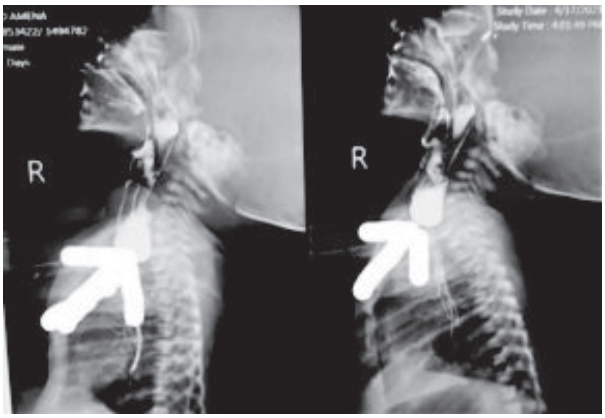

Fig.-2 : *Upper GIT contrast x-ray showing proximal dilatation of the oesophagus*

Case Discussion:

Oesophageal atresia is the most common oesophageal malformation in newborn. These patients can be diagnosed in a prenatal period, at delivery or in a postnatal period by the responsible physicians.[3] It was first described by Thomas Gibson in 1697. In case of oesophageal atresia (OA), oesophageal lumen is found to be interrupted, resulting in upper and lower segment[4]. Most of the patients (92%) usually have a trachea oesophageal fistula (TEF) and only 4% of patients with (TEF) do not have any OA.[5] This occurs only in 1/2500 to 1/4500 live births.[6,7] There is an alteration in the migration of lateral folds or growth arrests at the time of evagination. In most cases, TEF occurs due to incomplete separation of posterior oesophagus and trachea which occurs between third to sixth weeks of gestation.[8]

Anatomically, there are five subtypes of (Gross and Vogt) classification of OA on the basis of their relative frequencies. Type A: OA with distal TEF (86%). Type B: isolated OA (8%). Type C : isolated TEF (4%), Type D : OA with proximal TEF (1%) , Type E : OA with double TEF (1%)[9]. Our case was a rare type of oesophageal stenosis.

The antenatal diagnosis of OA/TEF can be done by USG scan which reveals polyhydramnios and proximal dilated blind ending oesophageal pouch.[10-11] The mother of our patient also had history of polyhydramnios. In the postnatal period, TEF can be suspected if there is excessive secretion per mouth, repeated episodes of coughing and checking, transient cyanosis shortly after birth. Inability to pass a rigid nasogastric tube can confirm the suspicion.[4] Our patient also had excessive secretion per mouth since1st day of life though we did not always face the difficulties during nasogastric tube insertion.

Mastroiacovo et al showed that, out of 92 malformation, 39 malformation including TEF/OA were more common in twins than singletons and in males.[12] In our case, the child was a male, singleton without any other dysmorphysms.

The association of other anomalies with TEF/OA is reported about 30-60%.[13] Most common anomalies are cardiovascular (11-49%) followed by genitourinary (24%), gastrointestinal (24%) and muskuloskeleal (13%).[14] Our patient had associated gastrointestinal malformation.

Some diagnostic modalities includes upper pouch oesophagogram (UPEG), tracheobronchoscopy and contrast oesophagogram with fluoroscopic control. However, oesophagogram with contrast barium studies offers the best visualization of TEF/OA.[15] We also performed an water soluble X-ray of upper GIT at the age of day twelve which revealed oesophageal stenosis.

Our patient was placed on oxygen, intravenous hydration, antibiotic therapy. Surgical management includes Neonatal Intensive Care Unit and appropriate anaesthesia. Prompt surgical intervention, ventilator support, prevention of septicemia should be ensured[15,16]. Unfortunately our patient was covid positive at 17th day of age and treated accordingly. So, the surgery was postponed. Surgery consists of ligation and division of the fistula with the repair of tracheal and oesophageal walls. Cervicotomy is preferred for proximal fistula and thoracotomy is preferred for distal fistula.[17] Brooke's et al reported that, Rt cervicotomy was done in 7 cases and Lt cervicotomy in 5 cases by Sundar et al[18,19]. On the other hand, Bhatnagar et al reported that, endoscopic treatment is more easier than open surgical procedures.[20] A fistula that has not closed after two endoscopic attempts should be approached externally.[21]

Conclusion:

Our case highlights that comprehensive clinical examination and meticulous follow up in newborn is essential in NICU set up. Besides these, ventilator support and other management modalities are mandatory in a tertiary hospital. All these efforts can prevent the high morbidity and mortality of newborns having TEF/OA. Besides these, risk of recurrence (1%) should be counselled about future pregnancy.

References:

1. Brzaèki V, Mladenoviæ B2, Jeremiæ L, •ivanoviæ D, Govedaroviæ N, Dragan Dimiæ D, Goluboviæ M et.al. Congenital esophageal stenosis: a rare malformation of the foregut. Nagoya J. Med. Sci. 2019, 81. 535–547
2. Singh AP, Ansari JS, Gupta P, Sharma P, Goyal RB. Congenital Esophageal Stenosis in an Infant. Journal of Evolution of Medical and Dental Sciences 2014; 3 (38), Page: 9820-9823.
3. Mutlu A, Kizilkaya O, Inan M. A Newborn with Osophageal Atresia, Tracheo-esophageal Fistula and feeding problems. Turkish Med Stud J 2019 :6(2);60-63.
4. Aminde L, Ebenye V, Arrey W, Takah N, Awungafac G. Oesophageal atresia and tracheo-oesophageal fistula in a preterm neonate in Limbe, Cameroon: Case report and brief literature review. Bio Med Central 2014: 7 :692.
5. Shaw-Smith C. Oesophageal atresia, tracheo-oesophageal fistula and the VACTERL association: review of genetics and epidemiology. J. Med. Genet 2006 , 43 : 545-554.
6. Garica H, Gutierrez MF. Multidisciplinary management of patients with Esophageal atresia. Bol Med Hosp Infant Mex 2011, 68(6) : 432-439.
7. Depaepe A, Dolk H, Lechat MF. The epidemiology of tracheo-oesophageal fistula and Oesophageal atresia in Europe. Eurocat Working Group. Arch. Dis Child. 1993, 68: 743-748.
8. Moore KL, Persaud TVN: Embryologia Clinica Mexico: McGraw-Hill interamericana;2001:288-289.

9. Vijayaraghavan BS: Antenatal diagnosis of oesophageal atresia with trachea-oesophageal fistula . J Ultrasound Med; 1996, 15: 417-419.

10. Pretorius DH, Drose JA, Dennis MA, Manchester DK, Manco JM. Tracheoesophageal fistula in utero . Twenty two cases. J Ultrasound Med; 1987,6: 509.

11. Malik L, Aziz S, MirzaS, Khan M. Unusual presentation of tracho-oesophageal fistula with Meconium aspiration syndrome in a preterm infant. Journal of Digestive disorders and diagnosis; 2017, 1: 8-12.

12. Haq A, Javeria S, IA Jan et al. Factors affecting survival in patients with oesphageal atresia and trachea-oesophageal fistula. J Ayub Med Coll Abbottabad ; 2009;21: 129-133.

13. Chittmittrapap S, Spitz L, Kiely EM, Brereton RJ. Oesophageal atresia and associated anomalies. Arch Dis Child, 1989; 64: 364-368.

14. Congenital anomalies , Walkers Paediatric Gastrointestinal disease 1: 14-19.

15. Al Rawi, Booker PD. Oesophageal atresia and tracho-oesophageal fistula. Contin Educ Anaesth Crit Care Pain 2007,7: 15-19.

16. Jaiswal A, Kumar A, Kumar M. 'H' type trachea-oesophageal fistula- Case reports with review of literature. Egyptian Journal of ENT and Allied Sciences, 2014, 15: 143-148.

17. Brookes JT, Smith MC, Smith RJ, Bauman NM, Manaligod JM, Sandler AD. H-type congenital trachea-oesophageal fistula : University of Iowa experience 1985 to 2005. Ann otol Rhinol Laryngol, 2007; 116 (5) : 363-368.

18. Sundar B, Guiney EJ, O'Donnell B. Congenital H-type tracheo-oesophageal fistula. Arch Dis Child, 1975; 50(11): 862-863.

19. Bhatnagar V, Lal R, Sriniwas M, Agarwala S, Mitra DK. Endoscopic treatment of tracheo-oesophageal fistula using electrocautery and the Nd: YAG laser . J Paediatr Surg. 1999; 34(3): 464-467.

20. Tzifa KT, Maxwell EL, Chait P et al. Endoscopic treatment of congenital H-type and recurrent tracheo-oesophageal fistula with electrocautery and histoacryl glue. Int J Paediatr Otorhinolaryngol ,2006;70(5): 925-930.

21. Vandenplas Y, Rudolph CD, DI Lorenzo, Hassal E, Liptak G, Manzur L. Pediatric gastroesophageal reflux clinical practice guidelines: joint recommendations of the North America Society for Paediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN)and the European society for Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN). J Paediatr Gastoenterol Nutr ,2009; 49: 498-547.