

Case Report

Esophageal Perforation in Neonates: a Report of Seven Cases from Kuwait

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ABSTRACT

Preterm neonates exposed to invasive procedures are at risk of esophageal perforation (EP). We are reporting a series of seven cases of EP, which have occurred in the neonatal intensive care unit (NICU) of Farwaniya Hospital, Kuwait, over a period of nine years (1994-2002). This gives an incidence of one in 8,118 live births and one in 278 NICU admissions. All patients were premature (below 32 weeks) and were exposed to various invasive procedures.

Two cases had a tentative diagnosis of esophageal atresia at the time of presentation. Other five cases presented

with a feeding tube in an ectopic position and air leak syndrome. All cases were managed conservatively. The perforations healed without any complication. The overall outcome was good as none of the mortality and morbidity was attributed to EP.

Our objective in this report is to highlight the existence of this problem and emphasize the importance of its early diagnosis, management and prevention. Misdiagnosis might lead to unnecessary investigation and / or surgical intervention.

KEY WORDS: congenital esophageal atresia, esophageal perforation, neonatal, NICU, preterm

INTRODUCTION

Esophageal Perforation (EP) is an iatrogenic condition, which occurs rarely in the Neonatal Intensive Care Unit (NICU). Most of the published literature reports one or two cases. Occasionally authors have published a series of cases collected over a number of years¹⁻⁴. However there has been no clear statement of the incidence of EP amongst the newborn population.

Krisna *et al*³ described the presentation of EP depending on the type of perforation. A Submucosal Esophageal Perforation (SEP) leads to a blind mediastinal fistula and mimics esophageal atresia or duplication^{3,4}. Free Esophageal Perforation (FEP) presents with a feeding tube in an aberrant position, pneumomediastinum, right-sided pneumothorax and/or pulmonary infiltration^{3,5}.

CASES REPORT

During a nine year period (1st January 1994 to 31st December 2002), a total of 56,827 live births were recorded in Farwaniya Hospital; 1947 of these were admitted to NICU. Seven cases of EP were diagnosed hence the incidence of EP is one in 8,118 live births and one in 278 NICU patients. The average occurrence of cases was at a rate of one case per two years, except for the year 1999, when two cases occurred over a five months period.

All patients were preterm with gestational ages ranging from 26 to 32 weeks. Their weight range was 650-1400 grams and five patients were small for gestational age (SGA). Five patients were male. The patient data are presented in Table 1. All patients were in the NICU at the time of diagnosis. They all had various exposures to invasive procedures: five patients were intubated during resuscitation, all cases were ventilated as a therapeutic intervention for hyaline membrane disease (HMD), all patients had soft sialistic oro-gastric tube (OGT) inserted on admission and changed regularly as indicated.

The time of presentation of EP was early, the first five days in six out of seven cases. Only case No. 1 presented at the age of ten days. Both cases of SEP presented very early, on the first day of life. The clinical signs were variable depending on the type of perforation. The five cases of FEP presented with right-sided pneumothorax or pneumomediastinum and/or abnormally positioned feeding tube (Fig.1). The two case of SEP (case Nos. 3 and 7) presented with bloody oral secretions and inability to pass the oro-gastric tube into the stomach. The lack of evidence of dilated esophageal pouch and absent coiling of OGT gave clues to the diagnosis (Fig. 2). Esophagogram, using Omnipaque 5 % radio-opaque dye, was performed on these two cases of SEP to confirm the diagnosis (Fig. 3: a, b)

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Table 1

Esophageal perforation in neonates: patient data

Clinical feature	Case	Case	Case	Case	Case	Case	Case
	1	2	3	4	5	6	7
Type of EP	FEP	FEP	SEP	FEP	FEP	FEP	SEP
Age of onset (days)	10	3	1	4	4	5	1
Gestational age (weeks)	28	32	30	27	26	29	29
Birth weight (grams)	1110	1400	932	680	740	650	950
Apgar scores at 1 & 5 min.	3, 5	5, 8	3, 7	2, 3	2, 4	4, 7	7, 8
Bloody oral secretions	-	-	+	-	-	-	+
Difficult passage of OGT	-	-	+	-	-	-	+
Rt. sided pneumothorax	+	-	-	+	+	+	+
Pneumo-mediastinum	-	+	-	+	-	-	+
Ectopic OGT in CXR	+	+	-	+	+	+	-
Abnormal contrast study	-	-	+	-	-	-	+

EP=Esophageal perforation, FEP=Free EP, SEP=Sub-mucosal EP
OGT=Oro-gastric tube, CXR= chest X-ray, + = present, - = absent

All patients were managed conservatively with bowel rest, nil per oral (NPO) for 7-14 days. Antimicrobial agents such as cefotaxime, amikacin, and metronidazole were given for one to two weeks. Nutritional and caloric requirements were maintained at a rate of 90-110 kcal/kg/day via total parental nutrition. All cases with pneumothoraces had a chest drain while pneumo-mediastinum was not drained and resolved spontaneously. Other supportive therapies such as ventilation, inotropic drugs etc., were given as indicated. Pediatric surgeon(s) were consulted in all cases. However, none of the patients needed surgical interventions during the course of illness.

The outcome was good in respect of EPs none of the mortality or morbidity was directly attributable to the perforation. Case No. 4 did not tolerate feeding and developed clinical and radiological signs of necrotizing enterocolitis (NEC) stage III-B. A peritoneal drain was placed but the poor general status of the patient prevented further surgical intervention. He died of fulminant peritonitis at the age of 32 days. Another patient (Case No. 3) died, aged-76-days, due to complications related to prematurity: post-hemorrhagic hydrocephalus and severe broncho-pulmonary dysplasia. The surviving patients were thriving well on out patient follow-up and had no long-term sequelae related to EP. Unfortunately case No. 5 was failing to thrive and had neurodevelopmental delay due to severe perinatal asphyxia.

DISCUSSION

Since Elkof *et al.* published the first report of EP in neonatal patients in 1969, many other worldwide reports followed^[6]. There have been occasional reports from the Middle East, but none from Kuwait until now^[7,8]. The incidence of EP amongst patients in our NICU is comparable to Grunebaum



Fig. 1: Case No. 1: chest X-ray showing right sided pneumothorax, pneumo-mediastinum and abnormally positioned OGT at base of right pleura



Fig. 2: Case No. 3: Chest X-ray showing pneumo-mediastinum and abnormally placed OGT in the mediastinum. No dilated esophageal pouch is seen.



Fig. 3: Case No. 7: Contrast study (CXR) showing the radio-opaque dye injected via OGT: a. Irregular spill out of radio-opaque dye into the upper mediastinum



b. Further tracking down of the dye into the lower mediastinum

et al as he reported three cases occurring in 787 NICU patients^[9]. The true incidence of EP might be much higher than reported in any neonatal unit as this condition could be asymptomatic or might present with non-specific signs, which could be misinterpreted as sepsis. Such cases would usually be managed conservatively with antimicrobial agents and by keeping them fasting^[10].

Premature infants exposed to invasive procedures such as endotracheal intubation, naso / oro-gastric intubation or oro-pharyngeal suction are a well-recognized risk group^[3,9,11]. EP has been reported to occur with various other invasive procedures such as chest drains, esophagoscopy, transesophageal echocardiogram, anti-reflux surgery or esophageal dilatation in cases of stricture^[5,8,12]. Although trauma is a major contributor to the etiology, spontaneous EP occurs in 17% of cases probably due to reflux of gastric contents against the closed upper esophageal end^[13,14]. Iatrogenic etiology is likely to be the contributor to EP in most of our patients, however the possibility of spontaneous EP does exist as premature infants have a high incidence of gastro- esophageal reflux and all our patients were premature^[15].

The clinical presenting signs depend on the type of perforation. The five cases of FEP presented with abnormalities in chest roentgenograms, right sided pneumothorax, pneumomediastinum and/or a feeding tube in an ectopic position. Although ventilated neonates are prone to baro-trauma which could manifest as pneumothorax or pneumo-mediastinum, the real clue to the diagnosis was the combined presence of an ectopic oro / naso-gastric tube with the evidence of air-leak. The explanation for the occurrence of pneumothorax, always on the right side, could be due to the protective role-played by the anatomical position of the aorta and the brachio-cephalic vessels in relation to the esophagus^[16]. The cases of

SEP (Case Nos. 3 and 7) presented with a clinical picture mimicking congenital esophageal atresia due to inability to pass the oro-gastric tube into the stomach. However, the presence of blood stained mouth secretions, radiological absence of dilated esophageal pouch and the absence of coiled OGT in chest roentgenogram gave clues to the diagnosis. Final confirmation was made by contrast studies and esophagogram (Fig. 3a, 3b). Although the diagnosis of EP can be confidently made on clinical and chest roentgenogram findings, many authors reported performing esophagograms to confirm diagnosis^[1,4,14]. Recently, ultrasound was used to locate the abnormal positioned OGT but this was not done in our cases^[17].

The management of EP in neonates, (contrary to the cases of EP in adults) as recommended in various reports, is conservative^[1,2,5,8,18,19]. All patients were treated conservatively by keeping them fasting for 10-14 days, administering broad spectrum antibiotics, metronidazole and total parental nutrition, in addition to supportive treatment as indicated. Leaving the mediastinal drain *in situ* was reported to have a controversial role in promoting healing^[18]. We have used it only in one case as suggested by a pediatric surgeon.

Surgical intervention is indicated in complicated cases such as unresolved pneumothorax, sepsis, mediastinitis or mediastinal collection as reported in previous studies^[1,20]. All cases of EP had an uneventful course of recovery. There were no early-related complications hence surgical intervention was not indicated. However, one patient had NEC, which has been reported as occurring in association with EP in two case studies^[1,20]. The question, which remains unanswered is whether this is a complication or an association. The possibility that it could be a co-incidental finding does stand true as both EP and NEC share common risk factors in sick premature infants. On follow up, none of the surviving patients had clinical evidence of esophageal stricture, which could be a long-term complication^[1,20].

In conclusion, a high index of suspicion is needed to diagnose EP in sick premature patients exposed to miscellaneous invasive procedures in the NICU. Conservative management with close observation for signs of complication will enable patients to avoid surgery, and will identify patients for whom surgical intervention is indicated. We hope that this report will raise the awareness of our neonatal colleagues about the possibility of this injury. They can avoid this complication by gentle and skillful action during newborn resuscitation, particularly in premature babies.

REFERENCES

- Bonnard A, Carricaburun E, Sapin E. Traumatic Pharyngoesophageal perforation in newborn infants. *Arch Pediatr* 1997; 4:737 - 743.
- Flores-Nava G, Joachin - Roy H, Rodriguez - Cueto G. Esophageal Perforation in premature newborn infants. Presentation of 8 cases. *Bol Med Hosp Infant Mex* 1993; 501:749-753.
- Krisna IH, Rosenfeild D, Benjamin BG, Klein G, Hiatt M, Hegyi T. Esophageal perforation in neonate: an emerging problem in the newborn nursery. *J Ped Surg* 1987; 22:784-790.
- Blair GK, Filler RM, Theodorescu D. Neonatal pharyngoesophageal perforation mimicking esophageal atresia : clues to diagnosis. *J Pediatr Surg* 1987; 22:770-774.
- Cairns PA, McClure BG, Halliday HL, Mc Reid M. Unusual site for oesophageal perforation in an extremely low birth weight infant. *Eur J Pediatr* 1999; 158:152-153.
- Elkof O, Lohr G, Okman L. Submucosal perforation of the esophagus in the neonate. *Acta Radiol* 1969; 8:187-192.
- Al-Khawahur H A, Al-Salem AH. Iatrogenic perforation of the esophagus Saudi Med J 2002; 23:732-734.
- Akel SR, Haddad FF, Hashim HA, Soubra MR, Mounla N. Traumatic injuries of the alimentary tract in children. *Pediatr Surg Int* 1998; 13:104 -107.
- Grunebaum M, Horodniceanu C, Wilunsky E, Reisner S. Iatrogenic transmural perforation of the esophagus in the preterm infant. *Clin Radiol* 1980; 31:257-261.
- Shah S, Coughtery H. Asymptomatic oesophageal perforation in neonate. *J Paediatr Child Health* 2001; 37:523-524.
- Seefeldes C, Elango S, Rosbe KW, Jennings RW. Oesophageal Perforation presenting as oesophageal atresia in a premature neonate following difficult intubation. *Paediatr Anaesth* 2001; 11:112-118.
- Muhiudeen-Russell IA, Miller-Hance WC, Silverman NH. Unrecognized esophageal perforation in a neonate during transesophageal echocardiography. *J Am Soc Echocardiogr* 2001; 14:747-749.
- Modi A, Mathur NB, Sarin YK. Spontaneous neonatal esophageal perforation. *Indian Pediatr* 2000; 37:901-903.
- Aaronson IA. Spontaneous rupture of esophagus in newborn. *J Pediatr Surg* 1975; 10:459-464.
- Byrne WJ. Fetal, transitional and neonatal physiology in: Schaffer and Avery's "Diseases of the newborn" 6th ed. Philadelphia: WB Sanders Co. 1991. p 663-664.
- Leonidas JC, Berdon W. The neonate and young infant: Pathologic conditions in the newborn Gastrointestinal Tract, in: Caffey's Pediatric X-ray diagnosis: volume 2, 9th ed. St. Louis: Mosby, 1993. p 2041-2043.
- Maruyama K, Shiojima T, Koizumi T. Sonographic detection of a malpositioned feeding tube causing esophageal perforation in a neonate. *J Clin Ultrasound* 2003; 31:108-110.
- Hager J, Menardi G. The diagnosis and therapy of esophageal perforation or rupture in infancy and childhood. *Rofu Fortschr Geb Rontgenstr Neven Verfah* 1990; 152:311-315.
- Johnson DE, Foker J, Munson DP, Nelson A, Athinarayanan P, Thompson TR. Management of esophageal and pharyngeal perforation in the newborn infant. *Pediatrics* 1982; 70:592-596.
- Sapin E, Gumpert L, Bonnard A, Carricaburu E, Sava E, Contencin P Helardot. Iatrogenic pharyngo-esophageal perforation in premature infants. *Eur J Pediatr Surg* 2000; 10:83-87.