Original Article

www.ijrhs.com
ISSN (o):2321–7251

Study of congenital diaphragmatic defects at a tertiary care teaching hospital in north coastal Andhra Pradesh

Venkata Ramana Poondla[1], Srilakshmi Kothakoona[2], Kameswari K[3], Janardana Rao K V[4]

1- Assistant Professor, Department of Paediatric Surgery, 2- Assistant Professor, Department of Anesthesiology, King George Hospital,Visakhapatnam, 3&4- Professor , Department of Paediatric Surgery, Andhra Medical College,Visakhapatnam, Andhra Pradesh, India

Submission Date: 19-01-2015 Acceptance Date: 27-01-2015

How to cite this article:

Vancouver/ICMJE Style
Poondla VR, Kothakoona S, KK, KV JR. Study of congenital diaphragmatic defects at a tertiary care teaching hospital in north coastal Andhra Pradesh. Int J Res Health Sci [Internet]. 2015;3(1):114-7. Available from http://www.ijrhs.com/issues.php?val=Volume3&iss=Issue1

Harvard style
Poondla, V.R., Kothakoona, S., K, K., K.V, J.R. Study of congenital diaphragmatic defects at a tertiary care teaching hospital in north coastal Andhra Pradesh. *Int J Res Health Sci*. [Online] 3(1). p.114-7 Available from: http://www.ijrhs.com/issues.php?val=Volume3&iss=Issue1

Corresponding Author:
Dr.Venkata Ramana Poondla, Assistant Professor, Department of Paediatric Surgery. Andhra Medical College, Visakhapatnam, Andhra Pradesh, India. Email: dr.pv.ms@gmail.com

Abstract:

Aim: To study the clinical presentation and outcome in Congenital Diaphragmatic defects. **Materials and Methods:** Retrospective study of 25 cases of Congenital Diaphragmatic defects including Congenital Diaphragmatic Hernia and Diaphragmatic Eventration admitted in King George Hospital of Andhra Medical College, Visakhapatnam, Andhra Pradesh from 2009 to 2014.Cases of age group from 1 day to 5 years are included in the present study. **Results:** 21 cases presented with classical clinical presentation, 2 cases with atypical presentation and 2 cases with post-operative recurrences which were operated outside the institution. Cases with Right side Diaphragmatic Hernia and a case with atypical presentation succumbed to death. **Conclusions:** Congenital Diaphragmatic Hernias are common on left side and carries good prognosis. Cases with atypical clinical presentation have 50% mortality. Right sided Congenital Diaphragmatic Hernias are rare but carries guarded prognosis.

Key words: Congenital Diaphragmatic Hernia, Diaphragmatic Eventration, Stomach Volvulus, Intestinal Malrotation.

Introduction:

Congenital Diaphragmatic defects embraces congenital diaphragmatic hernia and Diaphragmatic Eventration. It is one of the important causes for Respiratory distress in Newborn and infants, rarely with delayed presentation till childhood. It has an incidence of 1 in 5000 live births [1]. Advances in Radio Diagnosis facilitated early recognition of the condition in the fetus itself. The earliest English language description of the gross anatomy and Pathophysiology associated with congenital diaphragmatic hernia (CDH) in a newborn was by McCauley, an associate of Hunter, as reported in the Proceedings of the Royal College of Physicians, 1754. Until the 1980s, the standard of care remained immediate neonatal surgery followed by postoperative resuscitative therapy. It is essential to consider that the CDH is a physiologic emergency and not a surgical emergency.

Materials and Methods

25 children of Paediatric age group ranging from 1day to 5 years with diagnosis of Congenital Diaphragmatic Defects comprising Congenital Diaphragmatic Hernia and Diaphragmatic Eventration are taken up for study. These children were admitted during 2008 to 2014 i.e., over a period of 6 years. Diagnosis was arrived with clinical features, UltraSonography, Infantogram, Barium Meal Follow Through (BMFT) and CECT with oral contrast are the tools adopted for diagnosis. Repair of diaphragmatic hernia and Plication of Eventration of diaphragm under General Anaesthesia are adopted as treatment modalities. Assisted Ventilation provided for all the cases in the first 72 hours of post-operative period. The laterality of Diaphragmatic Hernia, Eventration of Diaphragm and their age distribution, outcome, complications and mortality are studied. 2 cases with atypical presentation and 2 cases with recurrence of Diaphragmatic Hernia, which were operated outside the institution, were also included in the present study.

Results

Of the 25 cases studied 21are male children and 4 are female at a ratio of 4:1.Majority of the children presented with classical clinical features in the neonatal period (17 cases), 5 cases are infants and 3 cases are of children in the age group of 1-3 year.3 of the 25 cases presented with atypical presentations like Volvulus of stomach (2 cases) and Malrotation of Gut (1 case).Left Diaphragmatic Hernia accounts to 16 cases and Diaphragmatic Eventration 9 cases. Left laterality is predominant in both Diaphragmatic Hernia and Eventration of Diaphragm, at a ratio of 21:4.No case of Eventration of Diaphragm on right side was reported. No case of bilateral Congenital Diaphragmatic Hernia was reported in the present study. Of the 5 children succumbed to death 2 are with left diaphragmatic hernia, 2 are with right diaphragmatic hernia and a case with left diaphragmatic Eventration. Overall prognosis is poor for right diaphragmatic hernia. 2 cases of Diaphragmatic Hernia with recurrence were repaired with success. No case of bilateral diaphragmatic hernia was recorded in the present study.

Table 1: Year and Gender wise distribution of cases

S.No	Year	Male	Female	Total
1	2008	2	0	2
2	2009	3	1	4
3	2010	4	0	4
4	2011	5	2	7
5	2012	3	0	3
6	2013	2	1	3
7	2014	2	0	2
	total	21	4	25

Table 2: Age wise Distribution of Cases

< 28 days	28th day – 1 year	>1 year
17	5	3

Table 3: Diagnosis and Laterality

S.No	Diaphragmatic Hernia		Total
	Left	Right	
1.	16	3	19

S.No	Eventration of Diaphragm		Total
	Left	Right	
1.	6	0	6

Table 4: Outcome and Laterality in Congenital

Congenital Diaphragmatic Hernia				Total
Good		Mortality		
Left	Right	Left	Right	
15	0	2	2	19

Eventration of Diaphragm				Total
Good		Mortality		
Left	Right	Left	Right	
5	0	1	0	6

Diaphragmatic Hernia and Eventration of Diaphragm

Figure 1: Showing Gastric Volvulus with Left Diaphragmatic Hernia

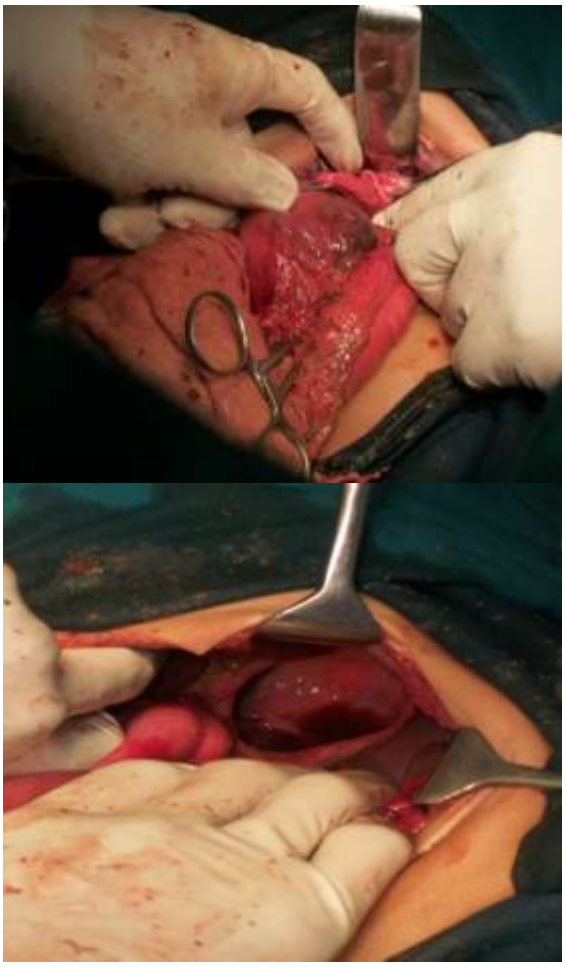
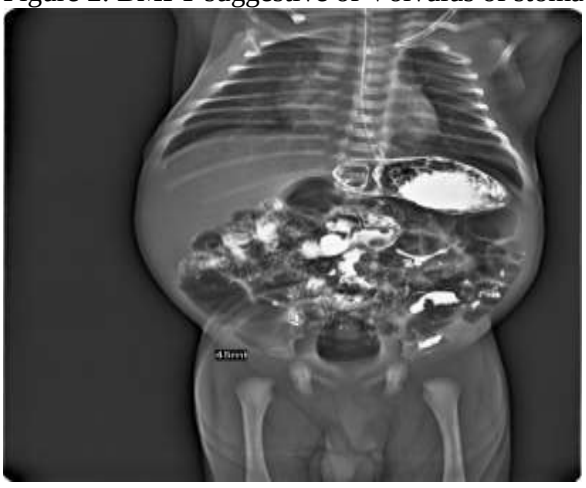


Figure 2: BMFT suggestive of Volvulus of stomach



Discussion

The incidence of associated malformations in infants with a CDH ranges from 10% to 50%. Skeletal defects have been noted in as many as 32% of CDH infants and include limb reduction and costovertebral defects. Cardiac anomalies have been found in 24% of infants including rare anomalies like Cantrell's pentology. In one study, 100% of stillborn infants with CDH had associated lethal anomalies. Even in infants who survive to birth but die shortly thereafter, neural tube defects were the most common malformations noted [2.3]. The diagnosis of a CDH is often made on a prenatal ultrasound (US) examination and is accurate in 40%to 90%of cases. Polyhydramnios has been reported present in up to 80% of pregnancies with associated CDH. Three-dimensional estimation of the fetal lung volume, calculation of the right lung area to thoracic area ratio, and calculation of the lung to thoracic circumference ratio are three different measurements that may correlate with neonatal outcome, but the lung-to-head ratio has been the most widely used prognostic indicator [4,5]. The most severely affected infants develop respiratory distress at birth, whereas a majority demonstrates respiratory symptoms within the first 24 hours of life. Although most CDH's present in the first 24 hours of life, 10% to 20% of the infants with this defect present later [6]. These latter infants present with recurrent mild respiratory illnesses, chronic pulmonary disease, pneumonia, effusion, empyema, or gastric Volvulus [Figure 1]. The respiratory distress associated with a CDH in the newborn results from a combination of two factors: uncorrectable pulmonary Hypoplasia and potentially reversible pulmonary hypertension. Atypical presentations of CDH with clinical features of Gastric Volvulus [7] [Figure 2] and Malrotation of Midgut [8] should be anticipated particularly in older children. Recurrent diaphragmatic hernia and small bowel obstruction are the dominant surgical challenges following initial repair. Recurrent hernias may occur in up to 50% of infants undergoing patch repair of the defect and in 10% of primary repairs, and they tend to occur in the first 4 years of life. Operative correction is easily performed through an upper transverse abdominal incision. Laparoscopic and thoracoscopic techniques [9] have also been used to repair this defect, but the laparoscopic approach is generally favored Even though Fetal Surgical Correction of CDH is described, the results are encouraging due to procedure associated maternal morbidity and mortality.

Conclusion

Even though majority of cases of diaphragmatic hernia and Eventration of Diaphragm presents with respiratory distress, Mediastinal shift, scaphoid abdomen and bowel sounds in the chest; atypical presentations like that of Malrotation of gut and Volvulus of stomach are common in delayed cases of congenital diaphragmatic defects.

Source of Funding: Nil
Source of Conflict: None

Acknowledgement

I am thankful to the Superintendent of King George Hospital, Visakhapatnam and the Principal, Andhra Medical College for extending their support for the present study. I also extend my regards to the parents of the children who co-operated me in the study.

Authors acknowledge the immense help received from the scholars whose articles are cited and included in references of this manuscript. The authors are also grateful to authors/editors/publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

References:

1. Congenital Diaphragmatic Hernia: Gallot D, Boda C, Ughetto S, Perthus I, Robert-Gnansia E, Francannet C, Laurichesse-Delmas H, Jani J, Coste K, Deprest J, et al: Prenatal detection and outcome of congenital diaphragmatic hernia: a French registry based study. Ultrasound Obstetric Gynecology 2007, 29:276-283. 18.

2. Sweed Y, Puri P: Congenital diaphragmatic hernia: influence of associated malformations on survival. Arch Dis Child 1993, 69:68-70.

3. Stoll C, Alembik Y, Dott B, Roth MP: Associated malformations in cases with congenital diaphragmatic hernia. Genet Couns 2008, 19:331-339.

4. Jani JC, Benachi A, Nicolaides KH, Allegaert K, Gratacos E, Mazkereth R, Matis J, Tibboel D, Van Heijst A, Storme L, et al: Prenatal prediction of neonatal morbidity in survivors with congenital diaphragmatic hernia: a multicenter study. Ultrasound Obstetric Gynecology 2009, 33:64-69.

5. J. Jani, R. L. Keller, A. Benachi et al., "Prenatal prediction of survival in isolated left-sided diaphragmatic hernia," Ultrasound in Obstetrics and Gynecology, vol. 27, no. 1, pp. 18–22, 2006.

6. Late-Presenting Congenital Diaphragmatic Hernia in Children: The Experience of Single Institution in Korea Dong Jin Kim and Jae Hee Chung, Yonsei Med J 54(5):1143-1148, 2013.

7. Congenital diaphragmatic hernia with gastric Volvulus, Prashant Jain, Beejal Sanghavi, Hemanshi Sanghani. S. V. Parelkar. S. S. Borwankar, Indian J. Surg. (December 2007) 69:260–263

8. Ford EG, Senac MO Jr, Srikanth MS, Weitzman JJ: Malrotation of the intestine in children. Ann Surg 1992, 215:172-178.

9. J Indian Assoc Pediatric Surg. 2010 Jan-Mar; 15(1): 2–8, minimally invasive surgery in neonates and infants Tiffany Lin and Ashwin Pimpalwar.