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Right Sided Congenital Diaphragmatic Hernia: A 10 Yrs Experience in a Tertiary Care Centre

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Abstract:
Aims: *To review the characteristics of patients admitted with right sided congenital diaphragmatic hernia (RCDH) and treated at a tertiary care centre over a period of 10 years.*
Materials and methods: *Records of patients diagnosed with RCDH who underwent treatment in a tertiary care centre between Jan 2009- Dec 2018. The variables noted down were: age at diagnosis, sex ratio, associated malformations, delay between initial presentation and diagnosis, clinical examination and operative findings.*
Results: *RCH constituted 15% of the 50 patients with CDH operated between 2009-2018. Mean age of diagnosis was 6 months(range 4 days- 16 months). Male to female ratio was ~ 1:2. Delay in diagnosis of symptomatic patients ranged between 0-12 months(mean 5.5 months). 2 patients presented immediately in neonatal period and 5 patients with delayed presentation (3 patients with milder symptoms and 2 patients with incidental diagnosis). Chest X ray(CXR) was diagnostic in 6 patients. Surgical repair by abdominal approach was possible in all 7 cases: suture repair in 6 cases and patch repair in 1 case. Hernial sac was found in 5 cases. Postoperative period was uneventful in all the patients.*
Conclusions : *RCDH is less symptomatic and milder in presentation as compared to LCDH. Majority of the cases could be diagnosed by CXR alone and all repaired by transabdominal approach. Hernial sac was found in 5 patients and correlated with milder and delayed presentation.*
Keywords: *Right congenital diaphragmatic hernia,CXR, hernial sac*

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I. Introduction

The term congenital diaphragmatic hernia (CDH) refers to the herniation of abdominal viscera through the posterolateral foremen of Bochdalek either to the left or to the right with an incidence of about 1 in 2500- 3000 live births . (1,2) RCDH accounts for 20-25% of CDH and is less discussed than the more common LCDH. (3,4) With milder and less acute symptoms, presentation beyond the neonatal period with atypical symptoms is not unusual in RCDH. So, the subtle differences in presentation between RCDH and LCDH should be known and one should have a high index of suspicion for its correct diagnosis.

II. Materials And Methods

Single centre retrospective study was carried out between January 2009- December 2018. Records of patients admitted with RCDH and subsequently treated were reviewed. The variables noted were age at diagnosis, sex ratio, associated malformations, the time lag between initial presentation and final diagnosis, clinical examination and operative findings.

III. Results

50 patients with CDH were operated between 2009-2018. 7 patients (15%) were found to have RCDH. Mean age of diagnosis was 6 months(range 4 days- 16 months). Male to female ratio was ~ 1:2. Age of diagnosis in the patients with delayed presentation ranged between 0-12 months(mean 5.5 months).
2 patients presented with acute symptoms in the neonatal period. 5 patients had delayed presentation : 3 patients with milder symptoms and 2 patients with incidental diagnosis while investigating for ASD.
2 patients had associated ASD whereas distal penile hypospadias was found in 1 case.
Chest X ray(CXR) was diagnostic in 6 patients. In one patient, it was interpreted as right lower lobe pneumonia but an ultrasound of the chest, prompted by a high degree of suspicion, settled the issue.
Surgical repair by abdominal approach was possible in all 7 cases: suture repair with polypropylene in 6 cases and patch repair with polypropylene- polyglactin mesh in 1 case with large defect.
Hernial sac was found in 5 cases.
Postoperative period was uneventful in all the patients.

IV. Discussion

The term congenital diaphragmatic hernia (CDH) refers to the herniation of abdominal viscera through the posterolateral foremen of Bochdalek either to the left or to the right with an incidence of about 1 in 2500-3000 live births . (1,2)

Embryologically diaphragm is derived from 4 sources(Kluth's hypothesis) but a 5 th source- post hepatic mesenchymal plate (PHMP) was postulated by Iritani in 1984.Diaphragmatic defect is clearly evident by 5 th week of gestation though the defect on the right occurs later (6th week) probably as a result of presence of inferior vena cava on the right side. (1)

RCDH accounts for 20-25% of CDH and is less discussed than the more common LCDH. (3,4) With milder and less acute symptoms, presentation beyond the neonatal period with atypical symptoms is not unusual in RCDH. Delayed presentation was noted in our series to the extent of ~ 90%. Osebold and Soper reported 5% of CDH patients presenting Rhonda the neonatal period but in other series the figures quoted were higher upto 25%. (5-7) Numanoglu et al reported 14% post neonatal symptomatic cases, one third on the right side. (8) Berman et al and Fotter et al found only 11% of delayed presenting CDH on the right side whereas Rasheed et al quoted the figure at 19%. (9-11)

Our results corroborated Berman's theory, which divides the patients into 2 groups: (a) group A where no hernial sac is detected and (b) group B with a hernial sac where the defect is obliterated by either the sac or the liver. Group A is the high risk group with acute presentation and Group B the low risk group in which the symptoms are usually delayed and insidious. (9)

The diagnosis of RCH can be made by a well performed CXR. (6,12) The classical features in CXR of RCH are a high right dome of diaphragm with hepatic flexors higher than normal. Berman et al reported 18% incidence of incorrect interpretation of CXR in LCDH. Two reports have mentioned the wrong interpretation of RCDH as pleural effusion. Studies of Berman et al, Blank and. Campbell, Fotter et al, Hartman and Numanoglu et al have stressed the well known possibility of a normal CXR done weeks or months before the diagnosis is made. (6,9,10) In our series, 80% of patients had delayed diagnosis due to incorrect interpretation of CXR.

Antenatal USG can diagnose CDH but may be reported as normal in the presence of hernial sac or the liver splinting the diaphragmatic defect. (6, 13)

As reported by various series by Berman et al, Delorimier, Hartman and Numanoglu et al , abdominal approach was preferred in our series as it comes useful in correction of rotational anomalies and has less morbidity than thoracotomy.

More well designed large multicentre prospective studies are required to make further conclusions.

V. Conclusion

RCDH is less symptomatic and milder in presentation as compared to LCDH delaying the presentation and diagnosis. Majority of the cases could be diagnosed by CXR alone and all repaired by transabdominal approach. Hernial sac was found in 5 patients and correlated with milder and delayed presentation. Successful repair was possible with abdominal approach with minimal morbidity.

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