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Congenital Diaphragmatic Hernia presenting after the Neonatal Period

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Summary

Adejuyigbe O, Abubakar AM, Sowande OA, Olayinka OS. Congenital Diaphragmatic Hernia presenting after the Neonatal Period. *Nigerian Journal of Paediatrics* **1998; 25:6.** Four cases of congenital diaphragmatic hernia (CDH), presenting after the neonatal period, at the Obafemi Awolowo University Teaching Hospitals Complex over a 12-year period (1983 - 1996), are reported. All the children were males, with a mean age of presentation, of six months (range, three - 11 months). Two of the CDH were through a right postero-lateral defect, one was through a left postero-lateral defect while the fourth was through a central defect in the diaphragm. The four children had persistent respiratory distress starting from birth and this was associated with pectus excavatum in the boy with the central diaphragmatic defect. Chest radiography and upper gastrointestinal contrast studies were diagnostic in three cases. Three of the four children had successful surgical repairs and although one of them later died from measles infection, the other two were in good health and thriving over a follow-up period ranging between three and 30 months. Our experience shows that CDH presenting after the neonatal period, poses less challenges in management if the diagnosis is made early and appropriate treatment instituted.

Inroduction

CONGENITAL diaphragmatic hernia (CDH) involves the herniation of the abdominal viscera into the thoracic cavity through a developmental defect in the diaphragm. Intrauterine lung growth and postnatal functions are compromised as a result of pressure effects.[1] Although it usually presents with respiratory distress at birth, and is a life-threatening emergency, some cases may remain undiagnosed until much later. [2-4] In this communication, we report four cases of CDH that presented after the neonatal period.

Patients and Methods

The case notes of four patients with CDH seen over a 13-year period (1983-1996) at the Obafemi Awolowo University Teaching Hospitals Complex, Ile Ife, were reviewed. Information extracted included age, sex, clinical presentations, results of radiographic investigations, findings at surgery and the outcome of treatment.

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Results

The children, who were all males, were aged from three months to 11 months at the time they first presented (Table 1). The presenting symptoms included difficulty in breathing since birth in all the four, failure to thrive in three and cough in two. Physical signs included tachypnoea, dyspnoea, unilateral diminution of breath sounds and the presence of bowel sounds over the thoracic cage. Only one of the four, a three-month old baby, who had features of heart failure, presented with cyanosis. Initial diagnoses included bronchopneumonia, CDH, congestive heart failure and eventration of the hernia. One of the four was initially thought to have bronchopneumonia with pyopneumothorax and was treated with tube thoracostomy. Chest radiographs revealed the presence of bowel loops in

three cases, and in one case, the radiographic changes were suggestive of eventration of the diaphragm. A more detailed summary of the clincal features and radiographic findings are contained in Table 1.

Table 1

Clincal Presentations of Four Cases of Congenital Diaphragmatic Hernia

Patients	*Age (Sex)*	*Symptoms*	*Signs*	*Results of investigations*
Case 1	6 months (M)	Difficulty in breathing since birth	Pectus excavatum	Chest X-ray': suggestive of eventration of the diaphragm
Case 2	3 months (M)	Difficulty in breathing since birth; cough	Dyspnoea. Dull percussion note: right hemithorax	Chest X-ray: presence of bowel loops in left hemithorax
Case 3	11 months (M)	Difficulty in breathing since birth; failure to thrive	Tachypnoea; apparent dextrocardia dull percussion note left hemithorax with reduced breath sounds; occasional bowel sounds in the left hemithorax; scaphoid abdomen.	Chest X-ray: shift of mediastinum to right and bowel loops in left hemithorax
Case 4	3 months (M)	Difficulty in breathing since birth; fever, cough; failure to thrive	Tachypnoea; cyanosis, dull percussion note right hemthorax, reduced breath sounds right hemithorax; tender hepatomegaly	Chest X-ray: ill defined diphragmatic silhoutte and bowel loops in right hemithorax; diagnosis confirmed by barium meal.

One of the children failed to report for surgery after the initial diagnosis, but showed up 16 months later. Unfortunately, he died immediately following admission and before surgery could be performed; the parents refused post mortem examination. The other three patients underwent successful surgery consisting mainly of reduction of the hernia and closure of defects. Details of the operative findings, the procedures carried out and outcome of surgery are shown in Table II.

Discussion

Congenital diaphragmatic hernia (CDH) still poses a major challenge of management. Despite improvements in prenatal diagnosis,[5] and *in-utero* tranfers to specialised centres for management, mortality is still very high.[2] The clinical course in an infant will depend on the degree of pulmonary hypoplasia present, the reversibility of the hypoplasia and associated pulmonary hypertension.[1]

Table II

Operative Findings, Surgical Procedures and Outcome of Treatment in Four Cases of CDH

Patients	*Operative Findings Site and Size of Defect*	*Contents of Hernia*	*Surgical Procedures*	*Outcome*
Case 1	Central defect to the right part of the diaphragm (10 x 6cm)	Part of the right lobe of the liver	Closure of defect with transversus abdominis muscle flap	Patient did well post-operatively but died of measles infection three months later
Case 2	-		None performed	Defaulted for 16 month. Died immediately on readmission before surgery. Parents objected to autopsy.
Case 3	Left postero-lateral (6 x 4cm)	Stomach, spleen, small bowel, caecum, appendix, ascending and transverse colon. Midgut malrotation with bands between loops	Reduction of hernia and closure of defect	Alive and well
Case 4	Right postero-lateral (8 x 6cm)	Small bowel, caecum, appendix, right half of transverse colon, right kidney and adrenal gland with a sac inverting over the kidney.	Reduction of hernia and closure of defect	Alive and well

CDH = Congenital Diaphragmatic Hernia.

The patients with CDH usually present at birth with respiratory distress. All our patiens presented 3-11 months after birth, although history showed that the symptoms started soon after birth, but were probably ignored or their importance missed by the caregivers. A review of the experience at Ibadan between 1970 and 1974, showed that two-thirds of their nine patients presented after the neonatal period while the remaining three patients presented within six days of birth.[4] A two-year old boy with CDH was also reported from Lagos in 1973.[4] We have not seen any case of the classic CDH at birth. This of course, does not indicate that such cases do not exist; they probably die shortly after birth, before gaining access to medical facilities. Furthermore, many of our women live in rural areas with no access to antenatal care and delivery by trained personnel. They are usually attended to, by traditional birth attendants who are ill-equipped to identify or cope with the challenges posed by CDH. Early feeding might also contribute to the early demise of such affected newborns.

In many centres, prenatal diagnosis by ultrasonography is the standard practice. Such babies are then transferred to centres where there are facilities for neonatal intensive care. In such centres, they undergo periods of preoperative stabilisation, well timed surgical repairs and aggressive post-operative intensive care.[1] In Nigeria, prenatal ultrasound evaluation is presently available on a limited basis.

Presentation of CDH afer the neonatal period is more varied and less dramatic than that in the neonatal period.[6] Some cases may be identified only at

autopsy of infants dying after sudden cardio-respiratory collapse.[7] Recurrent or persistent chest infections in a child should be viewed with suspicion of an underlying congenital intrathoracic abnormality. A high index of suspicion is also required in order not to miss the diagnosis in an older child. For instance, one of our patients was first diagnosed as having bronchopneumonia with pyopnemothorax and treated with tube theoracostomy before the correct diagnosis was made. This form of therapy could have worsened the respiratory function and increased the risk of perforating the bowel loops in the chest. The lesson to be learnt from this, is that suspicious chest radiographic findings should be clarified with a radiologist before a major therapeutic decision is taken.

The mortality rate for CDH is reported to be significantly lower in patients presenting after three days of birth than in those presenting earlier.[8] In this connection, it is pertinent to note that the three patients who had successful reduction of hernia contents and repair of their defects in the present series, did well post-operatively and although one of them later died, the death was unrelated to the CDH or its management. The remaining two patients are alive and well and are being followed up at our clinic. Despite the relatively good prognosis of CDH presenting after the neonatal period however, treatment should not be delayed once the diagnosis is made, even if the patient appears to be tolerating the condition well.

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