East African Medical Journal Vol. 81 No. 10 October 2004
NON-PULMONARY CAUSES OF ACUTE RESPIRATORY DISTRESS IN NEONATES: REPORT OF TWO CASES
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ABSTRACT

Acute respiratory distress (ARD) in neonates is a dangerous condition that is usually brought on by pulmonary dysfunction. Congenital diaphragmatic hernia and oesophageal atresia with tracheo-oesophageal fistula are examples of non-pulmonary causes of acute respiratory distress. Though rare, they are potentially fatal and successful management entails a multidisciplinary approach, which includes surgical intervention, use of critical care facilities such as the intensive care unit and paediatric supervision. We present two cases, each outlining one of the above named non-pulmonary causes of ARD, its management and outcome. Management of the congenital diaphragmatic hernia (CDH) was successful and the baby is still doing well. The baby with oesophageal atresia (OA) died during treatment, though the therapeutic procedures undertaken are presented here for academic purposes.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) and oesophageal atresia with tracheo-oesophageal fistula (TOF) is known to cause respiratory distress in neonates. Both conditions are uncommon and have an incidence of about 1 out of 4,000 live births(1). Differential diagnoses include pneumonia and/or meconeum aspiration. We present two cases that were managed in our health facilities in Mombasa, Kenya.

Case 1: Baby F. M. was born through a Caesarian section on 7th April 2001 to a 22 year old para 0+0 mother. Immediately after birth he was noticed to be distressed and cyanosed. He was resuscitated and improved on oxygen, but other episodes of cyanosis and distress re-occurred. Chest X-ray films (Figure 1) showed dextrocardia with consolidation of the right lung. The left lung had a mottled appearance and a large air bubble in the lower zone. Due to the recurrent episodes of distress he was transferred to intensive care unit (ICU) where he was intubated and ventilated. A nasogastric tube was passed. Subsequent X-ray films (Figure 2) showed tremendous improvement with good right lung aeration. This showed the left lung field remained mottled with gas shadows. The basal air bubble had progressively disappeared.

The abdominal ultrasound was only useful in reporting that the pancreas was not clearly visualized. A chest ultrasound was not done. Blood and urine tests were normal. Contrast studies X-ray films (Figure 3) were taken that suggested numerous loops of bowel in the entire left hemithorax. A firm diagnosis of diaphragmatic hernia was established and a decision to operate was made. A left parasternal (Kocher's) incision was made and the abdomen opened. A large diaphragmatic defect was found posteriorly (Bochdalek's hernia). Some of the abdominal contents (intestines, spleen and stomach) were found in the pleural cavity. There was no hernia sac. Reduction of hernia contents was done.

The hernia defect was repaired in two layers with No. 2/0 ethibond suture. A chest-tube was left in place connected to underwater seal drainage (Figure 4). The child did well post operatively and was discharged home within seven days. The lung eventually fully expanded (Figure 5). We have now followed this child for fourteen months and he is well.

Case 2: Baby A.T. was born on 18th May 2001 to a para 1+0 mother who had been antenatally investigated for hydromnios. At birth, he weighed 2.5 Kg. He was found to have respiratory distress with cyanosis. He was resuscitated and improved. He however, developed frequent cyanotic spells with coffee ground mucoid secretions from the mouth. He was transferred to ICU at The Aga Khan Hospital, Mombasa and put on a ventilator. A chest X-ray (Figure 6) was reported to show right side pneumonia with a large gastric bubble. An N/G tube could not pass beyond 10cm. The baby, however, remained pink with SPO2 of 100% while under ventilation. A little barium was introduced though the N/G tube (Figure 7) which confirmed oesophageal atresia. A diagnosis of oesophageal atresia with a tracheal-oesophageal fistula (TOF) was made.

Figure 1

A chest X-ray that shows mottled opacities of the left lung. The right lung is consolidated but this appearance may also be accentuated by the overlapping shadow of the right-sided heart. Note the large gastric air bubble at the left lower lung field

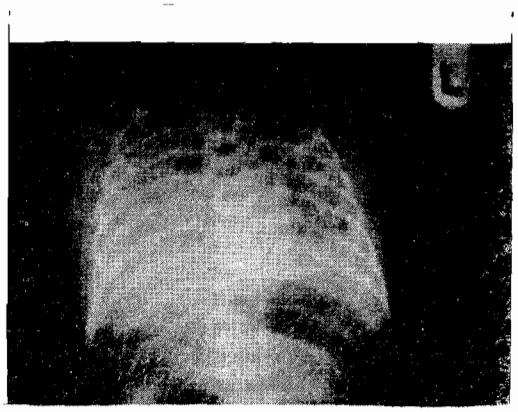


Figure 2

A chest X-ray after gastric decompression with a nasogastric tube

Figure 3

Contrast studies after introducing barium through the nasogastric tube. Note barium shadows in the left hemithorax

Figure 4

Immediate post-operative chest X-rays showing incomplete expansion of the left lung. Note the chest tube

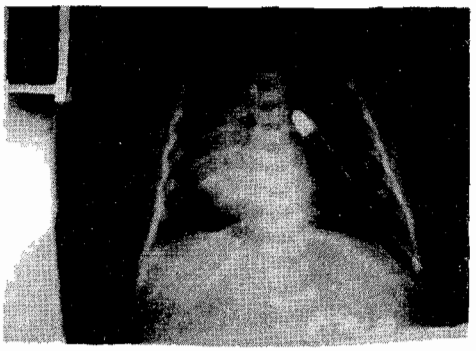


Figure 5

A chest X-ray showing better expanded left lung 10 days after surgery (compared to figure 4 above). Note also that the heart is in its normal position (left hemithorax). This confirms previous displacement (figure 1) by the abdominal contents has been corrected

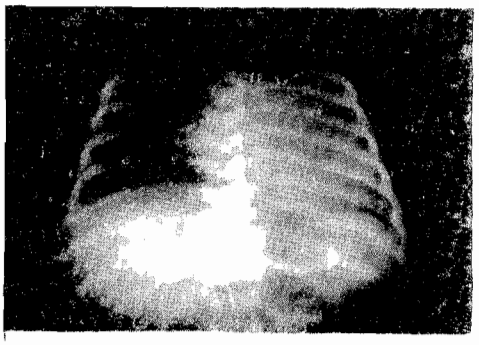


Figure 6

A chest X-ray film of a baby with TOF. Note the pneumonic process on the right upper zone and the large gastric bubble left lower zone

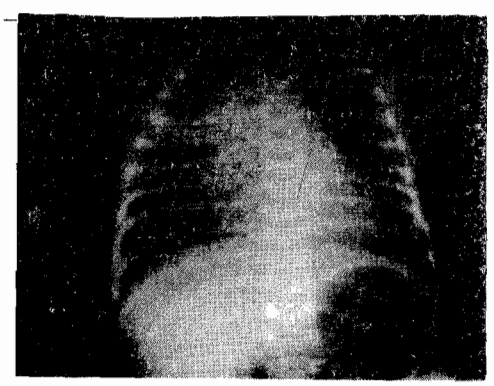
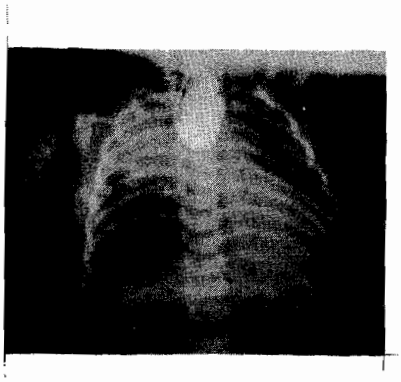


Figure 7

Introduction of small amount of contrast medium through the nasogastric tube to confirm the oesophageal atresia diagnosis. There is always the danger of ***aspiration*** *pneumonia during this process*



Exploration was done through a right side thoracotomy. A proximal atresia with a distal fistula was isolated and ligated. Primary anastomosis was done with 3/0 ethibond. A chest-tube was left *in-situ*. He did well in the immediate post-operative period. He remained pink with SPO2 ranging between 95-100%. He had total parenteral nutrition from the 2nd day. On the 6th post-operative day, he had a test feed (normal saline with methylene blue, a total of 60mls) with no demonstrable signs of leakage. On the seventh day the chest tube was removed. Several hours later a chest X-ray showed right lung collapse with mediastinal shift.

Another chest-tube was quickly inserted under local anaesthesia. A leak was suspected and confirmed with a dye fed through the N/G tube. The chest was re-explored and an anastomotic leak, measuring about 2x5 mm, was found anteriorly and was closed. The pleural cavity was cleaned and chest wall closed leaving in a chest tube. The child's condition changed at night three days later. He developed a fever, bradycardia and started to bleed through all the orifices. He died within a few hours.

DISCUSSION

The incidence of diaphragmatic hernia is 1:4000 in live births but increases to 1:2200 in still births(1). The defect is the absence of a portion of the diaphragm that leads to parts of the abdominal viscera entering the thoracic cavity. In most cases (90%), the defect is through the posterior part of the diaphragm, the foramen of the Bochdalek. Rarely (10%), it is due to failure of fusion of the central and lateral portions of the diaphragm anteriorly, leaving a defect known as the foramen of Morgagni. The latter rarely causes respiratory symptoms.

Children with Bochdalek hernia suffer respiratory distress in the immediate neonatal period as a result of pulmonary hypoplasia and compression of the pulmonary parenchyma by abdominal viscera (usually the spleen, stomach, left lobe of liver and the bulk of the intestines). This impairs the ipsilateral and sometimes the contralateral lung development. Severe hypoxia, hypercabia and acidosis then occur in the newborn. This gets worse after birth due to distension of the bowel by air entering the gastrointestinal tract, shifting the mediastinum and compressing the contralateral side. The babies then present with respiratory distress and cryamosis. The apex beat is displaced while the abdomen remains scaphoid. Breath sounds on the affected side are decreased while bowel sounds are rare.

Diagnosis is established radiologically, using a plain chest X-ray with the child upright that should show multiple loops of gut in the chest with loss of diaphragmatic outline. Eventration of diaphragm, pulmonary agenesis or hypoplasia and pneumatocoeles from staphylococcal pneumonia will give a similar radiographic pattern. Introduction of a small amount of contrast will unequivocally give the diagnosis. Success in the management revolves around use of critical care facilities and surgical repair. Mortality is high in most series: 80% in one series of 94 cases(2). The commonest cause of death is anastomotic leak and sepsis.

Oesophageal atresia with or without fistula is the commonest congenital anomaly of the oesophagus, occurring in 1 per 3000 to 4500 live births(3). Although the cause is mainly idiopathic, 50% of these babies have other congenital anomalies. In most cases, the mother has had polyhydramnios. Ninety per cent have some form of fistula the commonest being a proximal atresia with a distal fistula. The presence of fistula allows air from the trachea to the gastrointestinal tract causing abdominal distension. Any attempt to feed causes choking, cough and cyanosis. The baby may present with respiratory distress and cyanosis after birth.

Diagnosis is primarily established by passing a semi-rigid nasogastric tube through the mouth or nose that curls up in the proximal pouch. Erect radiographs that include the neck, chest and abdomen will show and determine the site of atresia. The presence of gastric and intestinal air on the abdominal film confirms the presence of a distal fistula (a small amount of water-soluble contrast may be used if there is doubt). The absence of air suggests a pure atresia without a fistula. The radiograph also shows associated cardiac or vertebral abnormalities and any pulmonary infection.

Management entails operative repair with ligation of fistula 2nd primary oesophageal anastomosis. Success in management will depend on the baby's weight, maturity, pulmonary status and the presence or absence of other congenital anomalies. Surgical repair is the definitive treatment.

We are presenting these two cases to highlight uncommon but treatable causes of respiratory distress in neonates. Both patients presented similarly but the radiological findings were as different as the structural abnormalities themselves. Aspiration pneumonitis is the more readily made diagnosis. A high index of suspicion is required followed by prudent investigations and consultations. The mortality rate decreases with the administration of critical care practices, parenteral nutrition and potent antibiotics. Most of these babies are operated in tertiary referral hospitals. The fact that the child with TOF died has not deterred us from sharing these interesting clinical cases.

ACKNOWLEDGEMENTS

To the Administrative Director, The Mombasa Hospital and the Medical Director, The Aga Khan Hospital Mombasa for allowing us to publish this study.

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