Congenital Diaphragmatic Hernia with Right Pneumothorax with Severe Lung Hypoplasia: Need for Gentle Resuscitation

Keywords: Diaphragmatic hernia; Pneumothorax; Gentle ventilation; Pulmonary hypoplasia; Pulmonary hypertension

Case

A term male baby with a birth weight of 2.8 kg was born to a primi mother with an Apgar score of 2/3/4. Baby was resuscitated after birth with bag and tube ventilation and shifted to the NICU on T piece resuscitation (Neopuff). Mother had a history of polyhydraminos and was diagnosed as a case of congenital diaphragmatic hernia antenatally. Baby was started on a ventilator and inotropic support. Initially baby was started on conventional ventilation and then shifted to high frequency ventilation as the baby was persistently hypoxic. Chest X-ray done showed left diaphragmatic hernia with mediastinal shift, severe lung hypoplasia and right pneumothorax (Figure 1,2). Baby Echo showed severe pulmonary hypertension. Baby expired at 6 hours of life secondary to severe lung hypoplasia and severe pulmonary hypertension.

Figure 1: showing left diaphragmatic hernia with severe lung hypoplasia, mediastinal shift and right pneumothorax.

Figure 2: shows left diaphragmatic hernia with severe lung hypoplasia, mediastinal shift and right pneumothorax.
Discussion

Congenital diaphragmatic hernia is herniation of abdominal content in thoracic cavity leading to lung hypoplasia and mediastinal shift [1]. This leads to severe lung hypoplasia and severe pulmonary hypertension, which is the major cause of mortality [2]. CDH is a rare condition that occurs in < 1-5:10000 births and left sided hernia are more than common than right sided hernia [3]. During resuscitation of antenatally diagnosed neonates with congenital diaphragmatic hernia, gentle ventilation must be done as the lungs are severely hypoplastic and frequently leads to pneumothorax [4]. CDH can be a component of syndromes which includes Pallister-Killian, Fryns, Ghersoni-Baruch, WAGR, Denys-Drash, Brachman-De Lange, Donnai-Barrow or Wolf-Hirschhorn syndromes [5].

Lesson to Clinicians

a. Resuscitation of these neonates must be gentle because of hypoplastic lungs as resuscitation with high pressure can lead to iatrogenic pneumothorax

b. Associated malformations must be searched in neonates of CDH as prognosis also depends upon associated features

c. Most common cause of death in CDH is pulmonary hypoplasia leading to pulmonary hypertension.

References