EDITORIAL COMMENT: CONGENITAL DIAPHRAGMATIC HERNIA

Inspite of many advances in neonatal surgery, the management of congenital diaphragmatic hernia through the posterolateral defect continues to pose a challenge to pediatricians and pediatric surgeons. Infants born with this anomaly, more often than not, have severe respiratory distress which demands prompt surgical reduction of hernia. The incidence of this anomaly in hospital births in India is reported as 1:1,200 (Hubli), 1:2,900 (Madras), 1:3,100 (Bombay, Delhi), and 1:5,800 (Madurai). Birth surveys in the West indicate an incidence of 1 in 5,000 to in 10,000 births. The frequency of associated anomalies in liveborn infants is very low indeed, but a high incidence of serious malformations has been reported in stillborn and aborted infants with congenital diaphragmatic hernia.

The main cause of death in untreated as well as operated cases of congenital diaphragmatic hernia is acute respiratory insufficiency caused by severe lung hypoplasia. Some infants die soon after birth, while others die due to respiratory distress that continues unabated following operation. A small number of infants remain undetected for several days and weeks and these have the best results following surgery. Nearly 80 per cent of the hernias of foramen of Bochdalek are on the left side. Right-sided hernia mostly contains only the liver. The prognosis of left-sided hernia depends on its size; smaller hernias having less severe symptoms and, therefore, better chances of survival.

It must be realised that the distension of herniated bowel is made worse by swallowed air, milk feeds and administration of oxygen under pressure with an open mask. Persistent distension also interferes with the venous return due to kinking and compression of inferior vena cava above the diaphragm. Severe restlessness due to hypoxia leads to hypoglycemia and hypothermia which aggravate the state of metabolic acidosis.

Early diagnosis depends on a high index of suspicion. If a newborn infant has dyspnea, tachypnea, seaphoid abdomen and dextrocardia, the diagnosis is congenital diaphragmatic hernia. Even the midwives and nurses should be taught to recognise these diagnostic symptoms. A skiagram of the abdomen and chest will show herniated loops of intestine displacing the compressed ipsilateral lung and the mediastinum to the opposite side and also interfering with the expansion of the contralateral lung as well.

As soon as the diagnosis is confirmed by radiographs, a wide bore (12 F) nasogastric tube is passed for continuous decompression of gastro-intestinal tract. Feeding is stopped and child is kept in a warm, humidified and oxygenated environment. Blood is collected for estimation of blood gases and for grouping and matching. An intravenous infusion line is established and systemic broad spectrum antibiotics are administered. Vitamin K is given by intramuscular injection. As soon as possible and preferably in the operation theatre, an endotracheal tube is passed and inter-
The Indian Journal of Pediatrics

158

mittent positive pressure respiration (I.P.P.R.) started using a gentle pressure of 15-20 cm of water. Temptation to over-inflate the compressed lungs must be resisted by the anaesthetist. Such a manoeuvre usually leads to pneumothorax, which may prove fatal. After taking the above measures, it is necessary to combat hypothermia, correct metabolic acidosis and improve blood glucose levels, as far as possible, before undertaking the surgical operation. It must be emphasized that a quick pre-operative correction of physiological imbalances mentioned above greatly improves the prognosis. If the blood gases return to normal after instituting the I.P.P.R. and metabolic acidosis is corrected with sodium bicarbonate solution (3 mEq/Kg) in 10 per cent glucose, the prognosis is usually good. If improvement does not occur, the abdomen should be opened without delay and visceras reduced into the abdominal cavity.

Repair of the diaphragmatic hernia can be easily achieved through a left or right subcostal incision. Some surgeons prefer a thoracic approach to a right-sided hernia. After reducing the hernial contents one should look for a hernial sac and recognise evagination of the diaphragm, if present. A gastrostomy is added to ensure effective decompression of stomach and intestines in the post-operative period. Usually the divided muscles are left ununited and only the skin is stitched. This intentionally-created incisional hernia provides the extra space needed to accommodate the replaced gastro-intestinal tract, spleen, liver etc.

Postoperative management ought to be carried out in an intensive care unit. Many surgeons institute under-water seal drainage of both the pleural cavities to ward off the dangers of pneumothorax, which is a dreaded complication of intermittent positive pressure ventilation.

Kerr has reported on lung functions at 7 to 19 years of age in 16 children after surgical repair of congenital diaphragmatic hernia. All were normal, except for episodes of minor wheezing in five. It seems that these had acquired disorders and there was no abnormality in lung function as revealed by clinical, radiological and respiratory function tests. It appears that hypoplastic lungs are capable of developing normally in due course of time.

It is true that some infants cannot be salvaged, mortality being due to severe pulmonary hypoplasia and irreversible hemodynamic changes. Research work carried out in experimental diaphragmatic hernia created in fetal lambs by intrauterine surgery has revealed the pathophysiology of respiratory insufficiency. The experimental lambs had pulmonary hypertension, severe patent ducts arteriosus, and low oxygen saturation in arterial blood similar to human infants. Right to left shunting can occur at the level of ductus, foramen ovale or within the hypoplastic unaerated lung. Resulting hypoxemia and acidosis perpetuate the vicious cycle, by causing pulmonary arteriolar vaso-constriction and pulmonary hypertension. Surgical ligation of ductus does not improve the clinical condition and the hypoplastic lung does not expand spontaneously after repair of hernia in lambs. Therapeutic agents, like tolazoline have been tried to lower the pulmonary pressure with some success. Newer research is now aiming at pharmacological control of pulmonary