ANAESTHETIC MANAGEMENT OF A PRETERM INFANT WITH PULMONARY HYPOPLASIA

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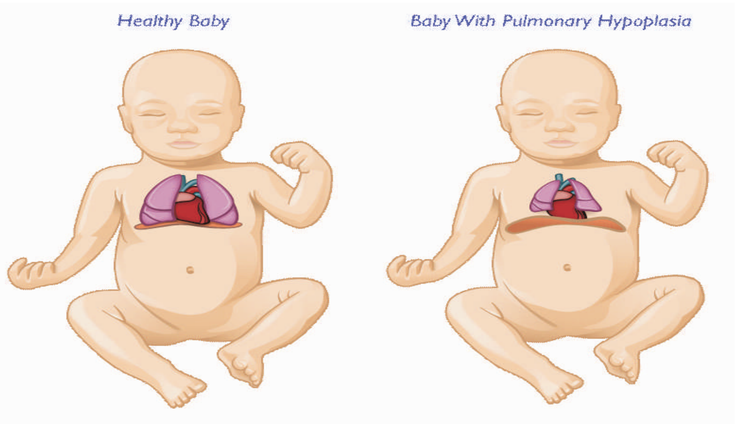


PULMONARY HYPOPLASIA- HISTORY

- A 2 month old male infant, a case of bilateral retinopathy posted for LSV
- Preterm born 26-28 weeks LSCS, elderly primi
- Birth weight 1.1 kilograms, Stormy neonatal period
- Ventilated for 70 days, O2 hood for 7 days.
- Treated sepsis, PRBC transfusion

PRE-OP EVALUATION

- Activity fair.When first seen, baby was on spontaneous ventilation with nasal O_2 of 3 liters. HR - 136 /min. Room air SaO_2 : 96-98%.
- CVS : S_1S_2 Normal,systolic murmur heard all over precordium
- Breath sounds normal on right side, uniformally decreased over left side.
- Haemogram - Normal
- CXR - left lung hypoplastic, right slightly hyperinflated.
- 128 slice CT pulmonary angio -

❖ left hypoplastic small pulmonary artery measuring 0.1cm with hypoplastic left lung.

❖ Normal cardiac situs with atrioventricular and ventriculoatrial concordance.

CT PULMONARY ANGIO...

❖ Pulmonary atresia with asymmetric bilateral pulmonary artery and small VSD and PFO.

❖ RA, LA, LV dilated.

❖ Left hypoplastic lung with crowding of ribs on left side.

❖ Right compensatory hyperinflation with herniation through the posterior junctional line to the left side.

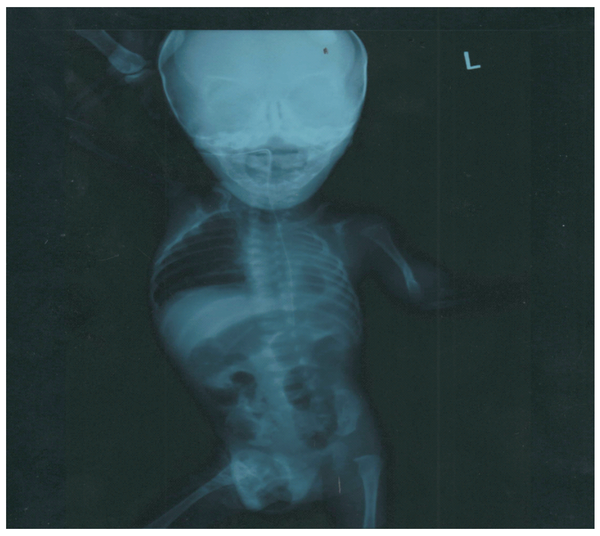
❖ Patchy enhancing area of consolidation with air bronchogram and adjacent fibrosis in the anterior and posterior segment of right upper lobe.

❖ Short segment 1 cm near total stenosis of left main bronchus.

- ECHO : Good bivent function , small VSD and PFO (L -R shunt).

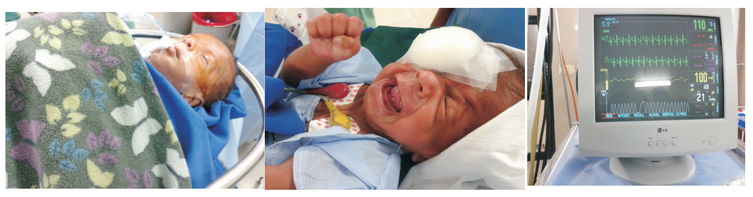
Mild PAH.

CXR SHOWING HYPOPLASTIC LUNG



PULMONARY HYPOPLASIA- HISTORY

- Premedication : 1/10th cc(0.01mcg) Pyrolate I.M. 45 minutes preoperatively.
- Monitoring - ECG, NIBP, SaO2, EtCO2
- Induction – O2, N2O(50 :50), Sevoflurane.
- Hot air warmer to maintain temparature
- I.V. access (24 G) after volatile induction
- VentIlation assissted using bag and mask
- After adequate depth, trachea intubated with 3.0mm plain ET tube in first attempt.
- Ventilation assissted manually with 2.5-3% Sevoflurane.



ANAEATHETIC MANAGEMENT

- Vitals stable intraop

HR - 120-130 /min

SaO2 - 100 %

EtCO2 – 20-25 mm of Hg

- I.V. Hydrocortisone 25 mg was given
- Total duration -two and half hours
- At the end of the procedure baby extubated after adequate respiration and good spontaneous activity.

DISCUSSION

- Pulmonary hypoplasia can be primary or secondary
- Pulmonary hypoplasia occurs commonly in association with CDH, Oligohydramnios (mostly relate to renal dysfunction), Skeletal dysplasias, Fetal hydrops, Malformation of CNS and Neuro muscular diseases .
- Secondary to other foetal abnormalities that interferes with normal development of lungs.
- Hypolastic lung - carina, malfomed bronchial stump, absent or poorly differentiated distal lung tissue.
- Small lungs - no normal gas exchange.
- Presentation is variable

Hsu JS, Lee YS, Lin CH, Li FY, Jeng MJ, Soong WJ, et al. Primary congenital pulmonary hypoplasia of a neonate. J Chin Med Assoc. 2012 Feb. 75(2): 87-90

- Normal lung growth requires

Normal - thoracic cavity , fetal breathing movements, amniotic fluid volume,

Fetal lung liquid at right pressure.

- Incidence - 9 – 11 /10,000 live births

14 /10,000 births

- Pulmonary Hypoplasia can be caused by:

❖ Abnormal thoracic cavity

Eg: CDH or malformations of chest cavity

❖ Abnormal fetal breathing movements- Neuro muscular disease (eg CNS lesions or SOL)in utero can result in decreased fetal breathing movements

❖ Abnormal amniotic fluid volume - due to renal agenesis, or urinary outflow obstruction or PROM

- For lung development to proceed normally, physical space in the fetal thorax must be adequate and amniotic fluid must be brought into the lung by fetal breathing movements, leading to distension of the developing lung.

(Intra thoracic cause of Pulmonary Hypoplasia)

DISCUSSION

- Several factors affect volume and pressure of amniotic fluid.
- Volume and Pressure-The volume of liquid in the lung is determined by the net rate at which liquid is secreted across pulmonary epithelium (4 – 5 ml/Kg/hr) and the rate at which it flows from the trachea into the fetal pharynx.
- The pressure in fetal trachea is normally about 2mm Hg higher than in the amniotic fluid thus preventing outflow of fetal lung fluid.
- Any alteration at 15- 28 weeks' of gestation can induce hypoplasia.
- Role of kidney in lung growth

Lung development starts in midtrimester with branching morphogenesis and is completed postnatally with the development of alveoli.

- Fetal urine is an important componant of amniotic fluid during late gestation and contributes to lung growth.
- During fetal development, kidney is also a major source of proline. Proline aids in the formation of collagen and mesenchyme in the lung, thus explaining severe hypoplasia in renal agenesis and dysplasias.

(Extra thoracic cause of Pulmonary Hypoplasia)

- Abnormalities of fetal lung fluid and lung fluid pressure - the underlying pathophysiology is unclear
- There can be overlap of aetiologies. Pulmonary hypoplasia may also be idiopathic or related to other syndromes and congenital anamolies- eg: multiple pterygium syndrome/ fetal akinesia-hypokinesia sequence(autosomal recessive)/Scimitar syndrome and trisomy 21.
- Congenital lung abnormalities are routinely detected at the time of routine high resolution prenatal ultrasound.
- Immediate presentation- difficulty in breathing with respiratory distress

Eg: cyanosis, intercostal recession with tachypnoea, acid base disturbance(acidosis, hypoxia,hypercarbia)

- There may be features of other fetal disorders eg skeletal dysplasias or Potter's facies in oligohydramnios.Pulmonary hypoplasia is suspected if oligohydramnios is present
- Other congenital anamolies eg cardiac malformations
- Fetal USG - can be used to measure lung area to head circumfrence ratio (a ratio of less than 1 is associated with high rate of neonatal death)
- Fetal chest circumfrence- reduced in case of intra thoracic causes.
- Doppler- peripheral pulmonary arterial resistance is often increased with pulmonary hypoplasia
- Fetal MRI - useful to calculate lung volumes. Fetuses with pulmonary hypoplasia will have low signal

Gerards FA, Twisk JW, Fetter WP, Wijnaendts LC, van Vugt JM. Predicting pulmonary hypoplasia with 2- or 3-dimensional ultrasonography in complicated pregnancies. Am J Obstet Gynecol. 2008 Jan. 198 (1): 140.e1-6.

Vergani P. Prenatal diagnosis of pulmonary hypoplasia. Curr Opin Obstet Gynecol. 2012 Mar. 24(2): 89-94

- *These babies are prone for chronic lung disease and have increased risk of morbidity and fatality from URTIs and LRTIs.*
- *Treatment: Antivirals, Antibiotics, Bronchodilators, inhaled corticosteroids (wheezing episodes)*
- *Mortality due to acute respiratory failure in the neonatal period, Chronic respiratory failure or insufficiency*
- *Pneumothorax either spontaneous or as a result of ventilatory support*
- *Persistant pulmonary hypertension caused by reduced pulmonary vascular bed and worsened by hypoxia or a coexisting L-R Intracardiac shunt.*
- *Chronic lung disease of infancy*
- *AsTLC is reduced - Prone for restrictive lung disease, recurrent respiratory infections, wheezing episodes, reduced exercise tolerance*
- *Scoliosis in adolescent years due to abnormal thoracic cage development*