新生兒持續性肺高壓

Pathophysiology

-failure to make the transition from high pulmonary vascular resistance(PVR) and low pulmonary blood flow to the relatively low PVR and high pulmonary blood flow of postnatal infant

Etiology

Pathology	associated disease	proposed mechanism	prognosis
Functional vasoconstriction, normal vascular development	acute perinatal asphyxia MAS. RDS sepsis or pneumonia hypoventilation, CNS depression, Hypothermia hypoglycemia	response to acute hypoxia particularly in the presence of acidemia	good, reversible
Fixed decreased Diameter Abnormal extension And hypertrophy of distal pulmonary vascular smooth muscle	placental insufficiency prolonged gestation *In utero* closure of ductus arteriosus(aspirin, NSAID) single ventricle without PS chronic pulmonary venous H/T (RAPVR, obstructive left-side heart disease)	Response to chronic hypoxia; Excessive pulmonary blood flow *in utero*, Elevated pulmonary venous pressure	poor, fixed structural lesion
Decreased cross-sectional area of the pulmonary vascular bed	space-occupying lesion (CDH, lung dysgensis, pleural effusion) congenital lung hypoplasia(Potter syndrome, thoracic dystrophies)	hypoplasia of alveoli and associated vessles	poor, fixed structural lesion
Functional obstruction to pulmonary blood flow	polycythemia hyperfibrinogenemia	increased blood viscosity	Good, unless chronic

Diagnosis

1. History
   - 甲、Prenatal history:IUGR, oligohydramnio, pleural effusion, maternal drug history(aspirin, indocid), space occupying lesion(CDH, lung cyst)
   - 乙、Birth history: asphyxia, MAS
   - 丙、Postnatal history: sepsis, pneumonia, airway obstruction
2. clinical presentation:
   - 甲、rapidly progressive cyanosis, tachypnea, respiratory distress, usually not compatible with the finding of the CXR
3. Examination
   - 甲、ABG: very low PO2 and SatO2 despite high FiO2;
     preductal PO2-postductal PO2≧15~20
   - 乙、Heart Echo:
     i. Firstly, cyanosis CHD must be ruled out
     ii. R to L(or bidirect) shunting at foramen ovale and/or PDA
     iii. TR with pressure gradient ≧ 40mmHg
     iv. Deviation of intra-atrial septum into the L’t atrium

丙、Distinguish with lung parenchymal disease and cyanotic congenial heart disease

i. Hyperoxia test: expose to 100% O2 for 5-10min→PO2 increase to<20mmHg

ii. Hyperventilation-hyperoxia test: expose to 100% O2 and RR 100~15-/min→PO2 increase at critical PCO2, often to <25mmHg

Assessing clinical severity

1. Alveolar-arterial oxygen gradient(AaDO2)
   甲、AaDO2= 760-47-(PaCO2/R)-PaO2
   乙、When FiO2=1.00→ R(呼吸商)≒1
   丙、If AaDO2>620→severe
2. Oxygen Index(O.I)
   甲、O.I= MAP*FiO2(%)÷postductal PaO2
   乙、MAP= mean airway pressure
   丙、If O.I >40→severe

Treatement

1. iNO
   甲、if iNO is available, it is the first choice
2. Alkalosis
   甲、Keep PH 7.5-7.55(reach this PH within 30-60min)
   乙、NaHCO3: about 1mEq/kg/hr; adjust by the PH of ABG
   丙、Taper: when PPHN improved ----1→0.5→0.25→0 mEq/kg/hr depend on the PH of ABG
   丁、F/U ABG, Na
3. HFOV
4. surfactant
   甲、indication: CXR showed RDS pattern and surfactant deficiency; PaO2<80mmHg under FiO2>40%
   乙、Dosage: 4cc/kg/dose
5. inotropic agent
   甲、indication: MBP<GA(preterm) or SBP<60mmHg(term)
   乙、Dobutrex: about 5-20ug/kg/min
   丙、Dopamine: keep urine output, about 1-2ug/kg/min; At RDS, try dopamine firstly, 5-0ug/kg/min
   丁、Epinephrine: 0.1ug/kg/min
6. correct underlying disease

7. supportive care
    甲、set IV, ETT, A-line(UA), UV as soon as possible, complete all procedures within 30-60min
    乙、correct hypoglycemia, hypocalcemia, systemic hypotension and acidosis
    丙、minimal handling
    丁、F/U ABG, Ca, Glu, Hgb, Na
8. Sedation
    甲、Not routine
    乙、Indication: irritable and fighting with ventilator while no other cause could be traced
    丙、: Drug: dormicum

Reference

1. neonatology, pathophysiology&management of the newborn, 5th ed, Avery
2. Avery's disease of the Newborn, 7th ed