



#### PROFESORES EXTRANJEROS

Vol. 30. Supl. 1, Abril-Junio 2007 pp S376-S384

# Post-operative respiratory failure after congenital heart surgery

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#### I. TIMING OF EXTUBATION

- a. Early Extubation
  - 1. Older infants and children
  - 2. ↓ complications (?): ↓ pulmonary issues, ↓ ICU length of stay, ↓ sedation, ↓ ↓ risk of pulmonary hypertensive crisis with suctioning
- b. Delayed extubation
  - 1. Hemodynamic instability
  - 2. Bleeding
  - 3. ECMO
  - 4. Open Chest
  - 5. Adequate pain control
  - 6. Infants < 4-6 months (?)

### **II. INCIDENCE OF FAILED EXTUBATIONS**

- a. Age Dependent
  - a. Infants at highest risk of failed extubation
  - b. Reported rates 11-27% in infants
- Risk factors: Age, genetic syndrome, circulatory arrest (stage I reconstruction), fluid overload, and pulmonary hypertension

## III. GENERAL MECHANISMS FOR RESPIRATORY FAILURE

- a. Pulmonary Diseases
  - 1. Intrathoracic Airway and Lung-↑ load
  - Alveolar disease (shunt and V/Q mismatch )-↑ shunt
- b. Extrathoracic Airway Abnormalities-- load
- c. Central and Peripheral Nervous System Disease-power
- d. Thoracic (skeletal structure and muscles) Diseasepower and/or - load

#### e. Treatment

- 1. Type I Respiratory Failure (Hypoxemia)-mean airway pressure and FiO<sub>2</sub>
- 2. Type II Respiratory Failure (Hypercapnia)-power/ load imbalance
  - vi. Relieve Airway obstruction
  - ii. Artificial Airway
  - iii. Mechanical ventilation

## IV. SPECIFIC MECHANISMS OF RESPIRATORY FAILURE IN PEDIATRIC CARDIAC PATIENTS

- Standard definitions may not apply and the usual therapies may not be appropriate
  - 1. Extubation may improve oxygenation/systemic oxygen delivery (i.e., TOF, Fontan, superior cavopulmonary anastamosis)
  - Hypoventilation to improve oxygenation/systemic oxygen delivery (i.e., bidirectional cavopulmonary anastamosis)
  - 3. Reducing FiO2 to improve systemic oxygen delivery (i.e., unrestrictive VSD, systemic to PA shunt)
  - 4. Extubatable settings does not mean the patient should be extubated (i.e., severe ventricular dysfunction)
- b. Remember CARDIOrespiratory failure (CARDIO is first and emphasized)!
- c. Pulmonary Edema
  - 1. Atrial/Pulmonary venous hypertension
    - i. Residual left to right shunts
    - ii. Diastolic dysfunction/Decreased ventricular compliance
    - iii. Systemic A-V valvular insufficiency/stenosis
    - iv. Outflow tract obstruction
    - v. Pulmonary vein stenosis/obstruction

- vi. Restrictive ASD in a functional single ventricle
- 2. Altered Alveolar Capillary Permeability
  - i. Transfusion Related Acute Lung Injury
  - ii. Cardiopulmonary bypass
  - iii.Drug reaction
- 3. Other
  - i. Mechanical during surgery
  - ii. Reperfusion injury
  - iii. Pulmonary Hemorrhage
- d. Acquired Lesions
  - 1. Extrathoracic airway
    - i. Edema- subglottic
    - ii. Vocal cord paralysis
    - iii.Subglottic stenosis/Granulation tissue/ Web
  - 2. Intrathoracic airway
    - i. Extrinsic compression
    - ii. Tracheobronchomalacia
    - iii.Stenosis/ granulation tissue
  - 3. Thoracic structures and musculature
    - i. Diaphragm paralysis
    - ii. Scoliosis
    - iii. Open sternum

- 4. Lung
  - i. Plerural Effusion
  - ii. Chronic lung disease
  - iii.Pneumothorax
- 5. Central Nervous System- stroke, hypoxia-, ischemia, seizures, hydrocephalus
- e. Congential Lesions
  - 1. Extra/Intra-thoracic airway
    - i. Genetic Syndromes (22q11 deletion, Trisomy 21, TOF, VACTERL, CHARGE, etc.)
    - ii. Absent Pulmonary Valve Syndrome
    - iii. Vascular rings (double Ao Arch, LPA sling)
  - 2. Congenital Lung Abnormalities
  - 3. Thoracic Structures and musculature
    - i. Diaphragmatic Eventration/Hernia
    - ii. Thoracic dystrophy
    - iii. Rib Abnormalities (VACTERL)
  - 4. Lung
    - i. Lobar emphysema
    - ii. Scimitar syndrome
    - iii. Hypoplasia (i.e., severe Ebstein's anomaly of TV)
  - 5. Congenital Neurologic Disease

#### V. CASE EXAMPLES

Case 1

A newborn with a prenatal diagnosis of Tetralogy of Fallot/ Absent Pulmonary Valve Syndrome. Did not require intubation after birth and had adequate arterial oxygen saturations after ductal closure. Initial CXR demonstrated a prominent MPA segment and mild hyperinflation of the right lung. An anterior plication of the MPA and LPA was performed to the hilum (LPA noted to be aneurysmal) and a valved RV-PA conduit was placed. Post-operatively there was hyperinlflation of the left lung and failure to wean from the ventilator (Figures 1 and 2).

After failed attempts at weaning from mechanical ventilation, a tracheostomy was performed. PEEP was used to the treat her tracheobronchomalacia with the best PEEP for maximizing expiratory flow rates at 12-14 cmH<sub>2</sub>O.



Figure 1.



Figure 2.

Case 2

A newborn with a pulmonary atresia/intact ventricular septum underwent a 4.0 mm right modified BT and required VA ECMO post-operatively after sudden cardiac arrest 24 hours after extubation. After weaning from ECMO support, he failed two trials of extubation and appeared to have poor

gas exchange with good respiratory effort. However, intubated there were no issues with gas exchange at low ventilator settings. Echo demonstrated good LV function, wide open ASD, no MR, no significant pericardial effusion and good shunt flow into the PAs (Figures 3 and 4).

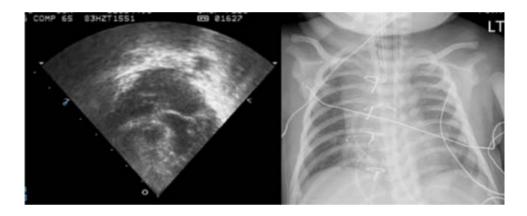


Figure 3.

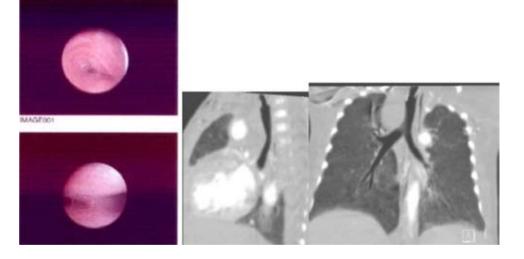


Figure 4.

A one -month old with an atrioventricular septal defect (AVSD) and left choanal atresia was transferred to CHOP for elective repair. He underwent a two patch repair of his AVSD and weaned from mechanical ventilation within 48 hours. He developed a mixed respiratory and metabolic acidosis and was reintubated. There was difficulty with

ventilation and abdominal distension with bradycardia during the intubation attempt. There were no issues with mask ventilation or intubation in the OR. After intubation and nasogastric decompression, his repeat X ray was back to baseline. He had poor respiratory system compliance (Figure 5).





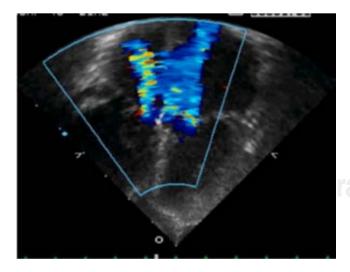




Figure 5.

An almost two week old presented to the emergency department with concern for worsening respiratory distress. He had previously been admitted overnight for similar complaints and had an airway fluoroscopy and upper GI study. On the day of this admission he presented with increased stridor, decreased activity and vomiting. He required intu-

bation after failing CPAP with an ABG of 6.99/114/92/27/-7. In the NICU there was difficulty ventilating the patient requiring the use of very high peak inspiratory pressures. The CICU was called to evaluate the patient since the patient also had congenital heart disease with a mid-muscular VSD and secundum ASD (Figure 6).

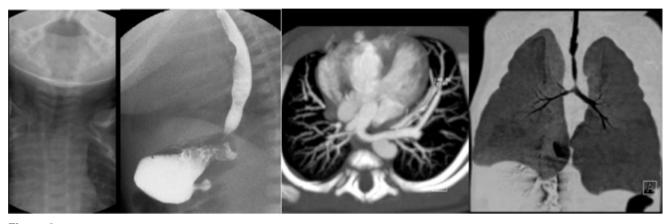


Figure 6.

#### Case 5

A newborn with a post-natal diagnosis of interrupted aortic arch type B and posterior malalignment type VSD underwent aortic arch reconstruction and Dacron patch closure of the VSD after recovering from her initial presentation in shock. Post-operatively she had adequate hemodynamics, but had to return to the OR for bleeding

shortly after arrival in the CICU. She was weaned from mechanical ventilation within 24 hours. She had adequate gas exchange after extubation. However, she was increasingly tachypneic. Her cardiac exam showed a soft systolic ejection type murmur, her perfusion was adequate, her liver was moderately enlarged (Figures 7 and 8).



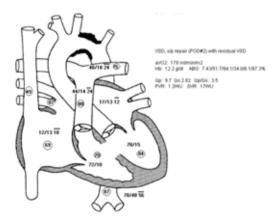


Figure 7.

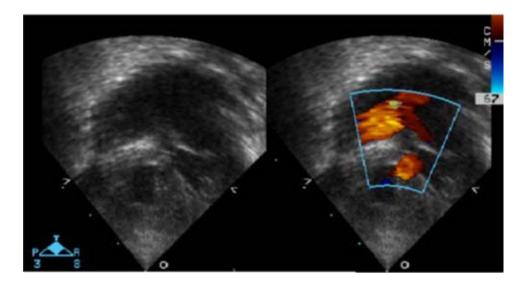


Figure 8.

Case 6

A 3 month Amish girl was admitted to the hospital for elective repair of her AVSD. Of note, on admission she was tachypneic with hepatomegaly and an elevated serum bicarbonate of 32 meq/dl. She underwent two-patch repair of her AVSD. Post-operatively she had poor urine output, tachycardia, and required higher ventilator infla-

tion pressures and set rate than the usual patient (rate of 30 bpm and PIP 30-35  $\rm cmH_2O$ ). Her echo showed good function mild MR, no TR, no pericardial effusion, and no residual VSD/ASD. CXR showed a bell shaped chest and pulmonary vascular congestion, but no other significant findings (Figure 9).

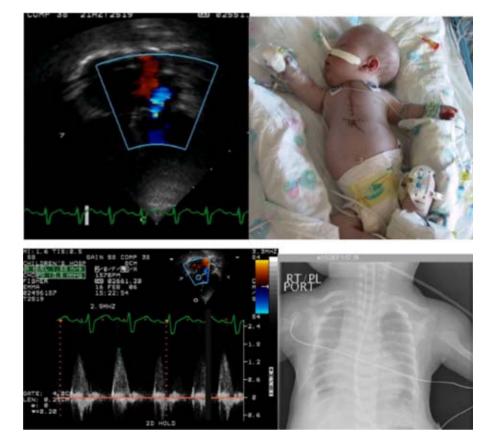
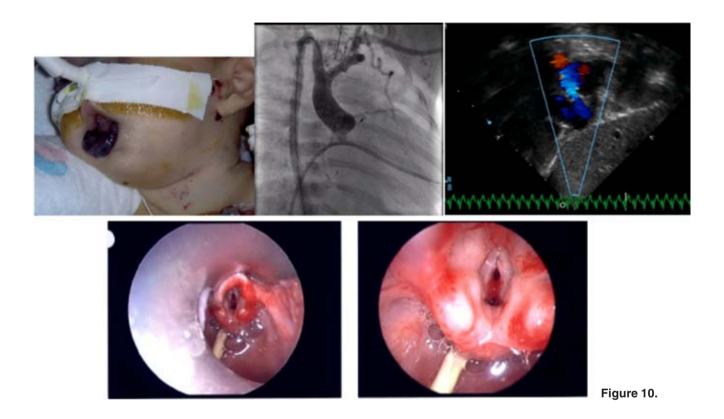


Figure 9.

A 2 month old with Tetralogy of Fallot, right aortic arch with severe coarctation s/p stent placement of the coarctation as a newborn presented for elective repair. Preoperatively she had mild-moderate respiratory distress. She underwent bronchoscopy, transannular patch with VSD closure

and jump graft from AAo to DAo. She underwent delayed sternal closure on POD #1 secondary to bleeding. She was noted to have hypoxemia while fully ventilated and muscle relaxed. She was given steroids and extubated on POD#3 (Figures 10 and 11).



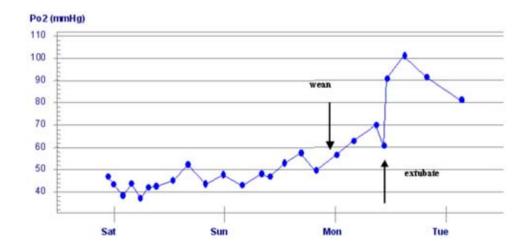


Figure 11.

A newborn female was prenatally noted to have pulmonary atresia with Ebstein's malformation of the TV, severe tricuspid regurgitation with dilated right ventricle and right atrium, and ductal-dependent pulmonary bloodflow. She was intubated after delivery secondary to respiratory distress. An operation was recommended for conversion to

single ventricle physiology. She underwent atrial septectomy, atrial reduction, plication of the RV w/pledgeted sutures, patch closure of TV w/4 mm fenestration and a 4 mm right modified BT shunt. She failed extubation within several hours on POD#3 secondary to progressive respiratory distress (Figures 12 and 13).

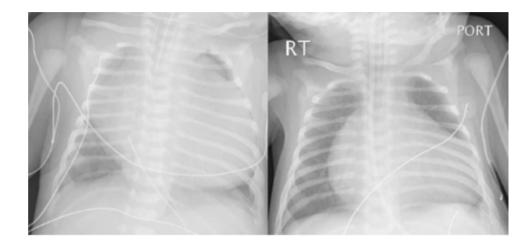


Figure 12.

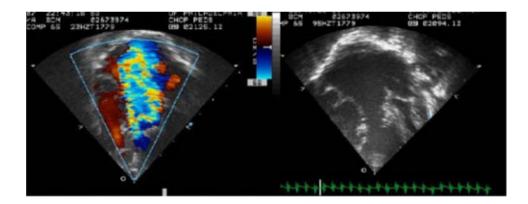


Figure 13.

#### Case 9

Fourth month old male, former 34 week premature infant with unbalanced common atrioventricular canal to the right and moderate- to -severe A-V valvular insufficiency had undergone a Stage I palliation with an RV-PA conduit. He also had a history of coarctation s/p Balloon dilation angioplasty  $\chi^2$ . He presented with a history of fever and decreased saturations. After an infectious etiology was ruled out, he underwent went a tricuspid valvuloplasty and bidirectional Glenn. He failed extu-

bation on arrival to the CICU secondary to profound hypoxemia. Despite reintubation and mechanical ventilation with 100% oxygen, he had pO<sub>2</sub>'s 2,5-30 mmHg. There was I-II/VI blowing systolic murmur at the lower sternal border and his liver was down 4 cm. He was warm peripherally with good perfusion. The RA pressure was 1,0-13 mmHg and with a good BP and mild sinus tachycardia. He was subsequently noted to rhinovirus positive (Figures 14 and 15).

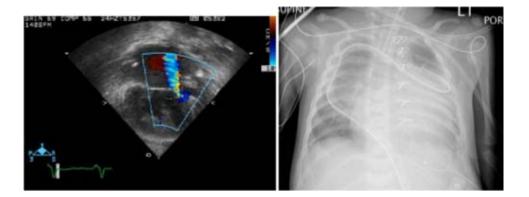


Figure 14.

