AGGRESSIVE RESPIRATORY THERAPY FOR CHRONIC PULMONARY DISEASE OPTIMIZES CLINICAL OUTCOMES FOLLOWING SURGERY FOR COMPLEX CONGENITAL HEART DEFECTS

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BACKGROUND

• Pulmonary Complications in children with complex congenital heart disease (CCHD) can have a profound adverse influence on the final outcome following surgery for CCHD.
• Early recognition and aggressive respiratory therapy has a critical role in the management of these patients.

METHODS

• A retrospective database search was performed to identify patients who attended both Cardiology and Respiratory Outpatient Clinics between January 1, 2010 and December 31, 2012.
• Entry criteria:
  1. CHD following surgery on home ventilations (O₂/CPAP/BiPAP);
  2. Prolonged ventilation > 14 days following initial surgery;
  3. Pleural effusion/chylothorax > 14 days following initial surgery.
• Respiratory therapy included the use of inhalers (bronchodilators and steroids), antibiotics and oral steroids.
• Clinical data including demographics, cardiac diagnosis, co-morbidities, surgical procedures, respiratory complications, respiratory and cardiac management, ejection fraction (EF) and pulmonary artery pressure (PAP) measurements, and pulmonary function tests (PFTs) were recorded.

RESULTS

• 24 children were identified, 56% male, median age 7.2 years (range 2-20).
• 54% presented with right sided lesions, 16/24 had multiple congenital anomalies.
• 40% met entry criteria of prolonged pleural/chylovus effusion or ventilation > 14 days post initial surgery.
• 18/24 had normal EF, median 64% (range 48-74).
• 21/24 had normal PAP (<25mmHg) with no significant RVOT obstruction (<20mmHg) indicating a satisfactory surgical result.
• 4/24 required cardiac transplantation.
• At the time of initial review 12/24 required some form of home ventilatory support despite normal EF and optimal cardiac repair; 9/12 had known upper airway lesions.
• At completion of the review period 8/24 were off ventilatory support.

CONCLUSIONS

• Despite obtaining an adequate mechanical repair with satisfactory ventricular function, significant respiratory dysfunction and sequelae from a complicated cardiac repair has a profound adverse influence on final clinical outcomes.
• Aggressive respiratory therapy can result in significant improvement in lung function and allow completion of multi-stage surgical procedures.
• In this series of patients, the lungs have a great capacity to heal.

Figure 1. (a) Pre-transplant. Patient with repaired DORV, tracheostomy, RPA stent in situ, coil/glue of occlusion of collaterals. Right pleural effusion; (b) Post-Transplant. Effusion resolved. Marked improvement in FEV₁/FVC.

Figure 2. (a) First x-ray. Patient with repaired TOF/DORV with prolonged chylous effusion. X-ray demonstrates persistent pleural effusion with interstitial thickening. Atelectasis in right upper lung; (b) Second x-ray. Two months post- cardiac transplant. Persistent rim of right pleural effusion, with atelectasis; (c) Third x-ray. One year post-transplant, symmetrical lung fields.

Figure 3. (a) First x-ray. Patient with dextrocardia and Fontan circulation. Second episode of plastic bronchitis, parainfluenza positive; (b) Second x-ray. Six months later following bronchoscopy and intensive antibiotic/steroid inhaler therapy.

Figure 4. Pre- and post-treatment pulmonary function results for patients able to complete testing.