Aggressive Respiratory Therapy for chronic pulmonary disease optimizes clinical outcomes following surgery for complex congenital heart defects.

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**Introduction.**

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

**Methods:**

A retrospective database search was performed to identify patients who attended both cardiology and respiratory outpatient clinics from January 2010 - December 2012.  
Entry criteria: 1)CCHD following surgery on home ventilation (Oxygen/CPAP/BiPAP); 2) prolonged ventilation >14days following initial surgery; 3) pleural effusion/chylous effusion >14days following initial surgery. Respiratory therapy included the use of inhalers (bronchodilators and steroid), 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 measurements (PAP), pulmonary function tests (PFTs) were recorded.

**Results**

24 children in total were identified, 56 % were male, median age 7.2 years (range 2-20yrs). 54% presented with right side lesions, 16/24 had multiple congenital anomalies. 40% met entry criteria of prolonged pleural/chylous effusion or ventilation >14 days post initial surgery. 18/24 had normal EF, mean 63%(range 48-74%). 21/24 had normal PAP (<25mmHg) with no significant RVOT obstruction (<20mmHg) indicating satisfactory surgical result. 6/24 required cardiac transplantation. Formal PFTs were obtained in 9/24 children (3 transplant). Pre-intensive respiratory therapy mean Forced Vital Capacity (FVC) was 47%(34-80%) predicted and Forced Vital Capacity (FVC) was 47% (FEV1 was 46% (16-86%). Following respiratory therapy both increased significantly, FVC 59% (21-82% p<0.001), FEV1 54% (16-81% p<0.03). At time of initial review 12/24 required some form of home ventilatory support despite normal EF and optimal cardiac repair; 8/12 had known upper airway lesions. At completion of the review period 4/8 were off ventilatory support.

**Conclusion**

Despite obtaining an adequate mechanical repair with satisfactory ventricular function, significant respiratory dysfunction as sequelae from a complicated cardiac repair has a profound adverse influence on final clinical outcome. Aggressive respiratory therapy can result in significant improvement in lung function (confirmed by PFTs) and allow completion of multistage surgical procedures. In this group of patients the lungs have a tremendous capacity to heal.