Plastic bronchitis; a potentially fatal complication after cavopulmonary connection such as Fontan and Glenn operations

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Plastic bronchitis (PB) is a rare and severe respiratory disorder, characterized by formation of gelatinous plugs bronchial casts in the large airways that take the shape of bronchial “casts.” Plastic bronchitis generally occurs in patients suffering from asthma, respiratory infections, cystic fibrosis, and lymphatic abnormalities; it is rarely potentially life-threatening complication of the Fontan operation, the final palliative procedure for various forms of single-ventricle heart diseases. PB is characterized by recurrent formation of large pale bronchial casts obstructing the tracheobronchial tree. The treatment includes inhaled or systemic steroids, aerosolized mucolytics, bronchosscopic lavage, direct bronchoscopic extraction and in few reports, aerosolized fibrinolytic such as inhaled plasminogen activator (rTPA) and pulmonary vasodilators. Hemodynamic alterations (elevated central venous pressure and low cardiac output) likely contribute to the formation of tracheobronchial casts composed of inflammatory debris, mucin, and fibrin. We treated 6 cases with PB, 5 Fontan and 1 Glenn procedures, two died, one underwent heart transplantation with no further episodes of PB, one successfully suspended rTPA after the implantation of a dual pace maker, one is successfully weaning rTPA with no recurrences. The exact causes of PB in these series of patients remain unknown. The optimization of hemodynamics, aggressive pulmonary vasodilatation, and inhaled t-PA is an effective treatment strategy for patients with PB after cavopulmonary connection. When the Fontan circulation fails or symptoms are unmanageable, PB may resolve after cardiac transplantation.

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Life time care for congenital heart patients: Demands, needs and difficulties faced by patients and profession

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95% of infants born with abnormalities of the cardiovascular system should reach adulthood, mainly due to surgical skills perfected over 60 years together with the development of pediatric cardiology, anesthesia, intervention, intensive care and refined imaging techniques. Best medical care and support must be continued beyond pediatrics. The needs of these grown up congenital heart patients [GUCHS] are many, medical for arrhythmias, heart failure endocarditis, symptomatic deterioration, increasing cyanosis, further cardiac surgery and/or intervention, surgery outside the heart and special care and advice in relation to pregnancy and disease in other systems. In addition, informed advice for life style issues and needs is required as part of specialized unit. The most “at risk” conditions are the named operations, any condition with atresia, transposition, malposition absence, single or common in title, childhood valve replacements and coarctation. Establishing best national medical care is difficult, obstructed by doctors, finance, management and “vested” interests. Overcoming them is possible and easiest in small countries with state funded health services.

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