

Pulmonary Hypertension in Congenital Heart Disease

-Congenital heart disease (CHD) is the most common congenital malformation and occurs in ~ 1% of all live births.

-Pulmonary hypertension may be present in patients with congenital heart disease (5-10% of all patients with CHD).

-Pulmonary hypertension in CHD patients is most often a result of defects that allow shunting of blood to the lung circulation, these defects may occur at multiple levels and vary in size and degree of shunt.

-Surgical or transcatheter correction of defects early in life may prevent long-standing pulmonary hypertension.

-Large central shunts that are not closed early in life result in pulmonary hypertension, elevation in pulmonary vascular resistance above systemic vascular resistance and shunt reversal, known as the Eisenmenger syndrome.

-Patients with the Eisenmenger syndrome are cyanotic (blue) because blue blood is shunted away from the lung arteries given the very high resistance to flow within these vessels.

-Patients with the Eisenmenger syndrome usually have limited exercise capacity.

-Patients with the Eisenmenger syndrome may survive into the 6th decade but are at risk for a number of adverse events, these include:

1. Bleeding (especially hemoptysis- coughing up blood).
2. Infective endocarditis
3. Arrhythmias
4. Heart failure
5. Pulmonary thrombosis (Clot in the lungs)
6. Paradoxical emboli (any clots or particles in the venous circulation can cross to the arterial circulation through the hole in the heart and cause problems- e.g. stroke).

-Fortunately, many of the same therapies now used in other pulmonary hypertension patients may improve functional capacity in patients with CHD. The ones that have been tested in CHD thus far include: Bosentan, sildenafil, tadalafil, and Flolan.

