D-TRANSPOSITION OF THE GREAT ARTERIES

Note: "D" *denotes* ***dextroposition*** *of aorta, i.e. it is anterior and to the right in relation to the pulmonary artery. TGA accounts* ***≈5%*** *of all CHD,* ***Male>****Female; it also more common in patients with CATCH 22 (or DiGeorge synd) & infants of diabetic mothers. The survival in these newborns is depends on the* ***foramen ovale*** *and the* ***ductus arteriosus****.*

D-TGA with Intact Ventricular Septum

(Simple or Isolated TGA) *Before birth, oxygenation of the* ***fetus*** *is near* ***normal****, but after birth, once the* ***ductus begins to close****, the minimal mixing of systemic and*

pulmonary blood via the ***patent foramen ovale become insufficient***

and severe hypoxemia ensues, generally within 1st few days of life. ***C.M. Cyanosis and tachypnea*** *usually become evident within the* ***1st hours or days*** *of life. It is a medical emergency because if untreated, the*

vast majority would not survive beyond the neonatal period due to ***severe hypoxemia & acidosis****, but HF is less common. Other clinical findings may be* ***subtle*** *& nonspecific, even the murmur*

may be ***absent*** *or there is* ***soft systolic ejection murmur****.*

Inv. *□* ***CXR;*** *may be normal or show mild cardiomegaly & narrow mediastinum* ***"Egg-shaped heart"*** *with normal or ↑ pulmonary blood flow. □* ***ECG;*** *shows the normal neonatal right axis deviation. □* ***Echo, Doppler & Ventriculography*** *studies are diagnostics. □* ***Catheterization*** *may be done in inconclusive cases or when the patient require emergency balloon atrial septostomy.* ***Note:*** *Anomalous coronary arteries are noted in 10–15% of patients.* ***Rx.*** *When TGA is suspected, an infusion of* ***PGE1*** *should be initiated*

immediately. Patient should also kept ***warm*** *with correction of* ***acidosis & hypoglycemia****.*

Rashkind balloon atrial septostomy *can be done if there is severe hypoxia or acidosis despite prostaglandin infusion, or there is any significant delay in corrective surgery. Rashkind septostomy may be done even if VSD is present (especially if there is poor mixing of blood).*

*Corrective procedure of choice is the **Arterial Switch** (Jantene*

*procedure) performed within the **1st 2 wk** of life because pulmonary vascular resistance declines after birth resulting in ↓ of the left ventricular mass, therefore, if this operation is delayed, the L.V. cannot pump blood to the systemic circulation. **Atrial Switch procedure** (Mustard or Senning operation) by intra-atrial*

baffle; however it has many Cxs, thus it is only reserved in cases with complex anatomy.

D-TGA with VSD

*If the VSD is **small**, the manifestation are **similar** to those of **intact***

ventricular septum (even no need to be closed during surgery), whereas

***large** VSD which is non-restrictive, i.e. causing a significant **mixing** of oxygenated and deoxygenated blood, may result in features of **HF** & the*

***cyanosis will be subtle or delayed** for several months. **Inv.** □ **CXR;** cardiomegaly, narrow mediastinum, and ↑ pulmonary vascularity. □ **ECG;** R.V. or biventricular hypertrophy. □ **Echo & Ventriculography** are diagnostics. □ **Catheterization;** peak systolic pressure is equal in the 2 ventricles & the 2 major arteries (aorta & pulmonary). **Rx. Surgery** is consist of arterial switch procedure with VSD closure (if large) within the 1st months of life, before the development of HF, pulmonary hypertension, & FTT..*