Introduction

Congenital heart disease (CHD) is one of the most common birth defects, affecting around 1% of live births ***(Lewin, 2000)***.

Both environmental factors and genetic factors have been implicated ***(Goldmuntz, 2001)***. Ocular studies in CHD are few and have concentrated on one cardiac anomaly,one syndrome with cardiac anomaly or single case reports. ***(Megarbane et al., 2000 & Twelker and Mutti, 2001)***.

The large percentage of ocular findings in CHD could be related to the high incidence of associated syndromes, to the possible embryologic link between the ocular and cardiac defect, or to the high incidence of consanguinity ***(Mansour et al., 2002 & Beratis et al., 2000)***.

Retinal vascular tortuosity is related to oxygen saturation, children with cyanotic congenital heart disease reportedly have increased retinal vascular tortuosity, retinal hemorrhages, and papilledema believed to be related to hypoxia and erythrocytosis ***(Tornqvist et al., 2002 & Wirth, 2002)***.

Hypoxemia, whether induced at high altitude or due to cyanotic congenital heart disease, results in a marked upregulation of angiogenesis-related vascular endothelial growth factor (VEGF), stimulating abnormal blood vessels formation (neovascularization), children with cyanotic heart disease have elevated systemic levels of

VEGF and increased retinal tortuosity ***(Ootaki et al., 2003& Pokharel et al., 2000)***.

Diller et al., 2006 proposed that hypoxia as well as erythrocytosis plays a key role in the retinal vascular patterns of children with cyanotic congenital heart disease.

Pulsatile retinal arteriolar tortuosity has been previously reported in 50% of patients with coarctation of aorta ***(Mayer et al., 2001)***. However, more recently, children with coarctation of aorta have been found not to display these findings because of early surgical correction of the cardiac lesion, implying hemodynamic aetiology of vascular tortuosity ***(Dimopoulos et al., 2008)***.