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# Coats' Disease in a Patient with Anterior Segment Dysgenesis

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**Abstract:** This paper describes a rare case of Coats' disease in a child with underlying Anterior Segment Dysgenesis (ASD) with secondary glaucoma in the contralateral eye.
**Keywords:** Coats' Disease, Anterior Segment Dysgenesis, Secondary glaucoma

## INTRODUCTION

We present a rare case of Coats disease which was seen in a child who has underlying Anterior Segment Dysgenesis (ASD) with secondary glaucoma in the contralateral eye. From our readings, this is the first case with dual different pathological ocular problems in a child being reported.

## CASE HISTORY

A day 5 of life child with underlying Tetralogy of Fallot presented with bilateral cornea opacity since birth. The child was born to non-consanguineous parents after a normal pregnancy and delivery. At presentation, the right eye blinked to bright light but was not fixating. The left eye had steady and maintained fixation to light. There was a dense cornea opacity in the right eye, with no view of the iris, pupil and fundus. The left cornea was clearer with good view of iris details. Dilated fundus examination showed non-glaucomatous optic disc and normal retinal vasculature.

Due to his cardiac status and the unavailability of paediatric post-operative intensive care, examination under anaesthesia (EUA) was only performed at 8 months of age. Examination showed dense central cornea opacity over the right eye with irido-corneal touch (Figure 1). B scan showed clear vitreous and flat retina (Figure 2). The corneal diameter of the right eye was 12mm horizontally. The left eye cornea was clear with no significant ocular abnormality. Intraocular pressure with Perkins Tonometer was 22 mmHg on the right and 10 mmHg over the left eye. A diagnosis of right eye anterior segment dysgenesis with secondary glaucoma was made and Baerveldt Glaucoma Drainage Device was implanted. Optical iridectomy was performed in that eye.

The Baerveldt implant was explanted after 6 months as it was found broken and the corneal diameter has increased to 14mm horizontally. Cyclocryotheraphy was done to the right eye. The intraocular pressure subsequently remained controlled with no further increment in corneal diameter and the child was asymptomatic.

A year later the vision deteriorated as the mother noted the child had frequent episodes of falls. Vision assessment showed the right eye blinked to light, but the left eye could not perceive light. Ocular examination under anaesthesia revealed telangiectatic vessels (Figure 3) with haemorrhage and exudative retinal detachment over the left eye (Figure 4 and 5). Intraocular pressure was normal. Refraction over the left eye showed significant hypermetropia +18.0DS/-1.50DC x 180. A diagnosis of Coats' Disease was made. Intravitreal Ranibizumab 0.5mg in 0.05ml was given with periocular steroids (triamcinolone 20mg and Dexamethasone 4mg) to reduce the exudation for retinal ablation of the telangiectatic vessels.

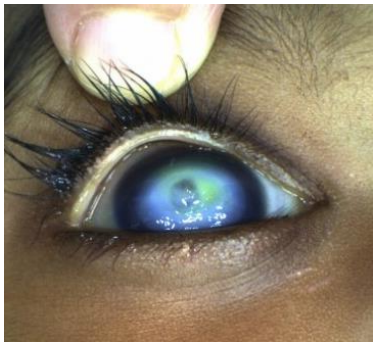

**Fig-1: Right eye cornea opacity**

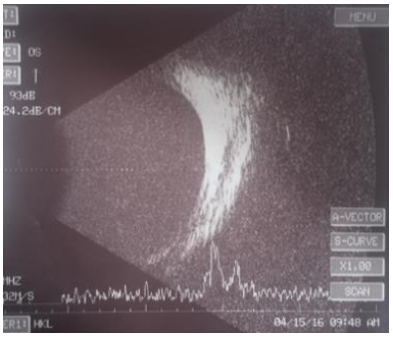


**Fig-2: Right eye B scan shows clear vitreous with flat Retina**

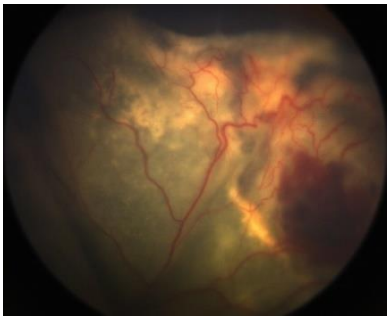

**Fig-3: Left eye shows telangiectatic vessels and exudation**

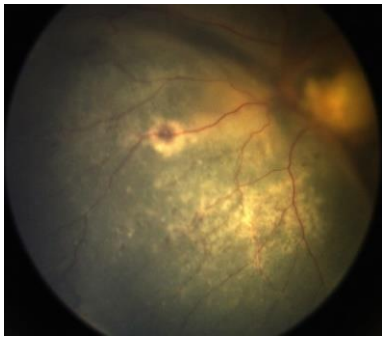

**Fig4: Left eye exudative retinal detachment**

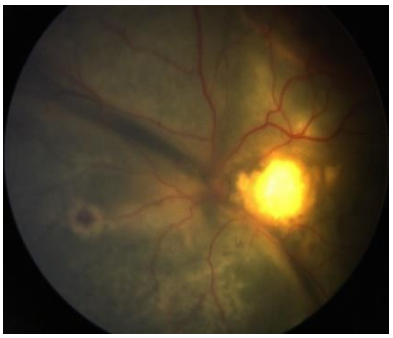

**Fig-5: Total left eye exudative retinal detachment**

## DISCUSSION

Anterior segment dysgenesis (ASD) is a spectrum of developmental conditions affecting the cornea, iris and lens. Patients with ASD often have malformations of the tissues responsible for intraocular pressure (IOP) regulation and aqueous humour drainage. They frequently develop elevated IOP, putting them at risk for developing glaucoma. It is generally associated with an approximate 50% risk for glaucoma.

To the best of our knowledge, this is the first report of Coats' disease observed in a child with underlying unilateral anterior segment dysgenesis. The association of anterior segment dysgenesis and coats' disease are not fully understood. However, recent studies showed retinal vascular abnormalities and peripheral nonperfusion was found to be related with childhood glaucoma [1]. Fundus fluorescein angiogram showed retinal vascular nonperfusion in patients with both primary and secondary childhood glaucoma [1]. Other common findings include circumferential branching of the vasculature parallel to ora serrata, anastomotic vessels and capillary dilatation. Leakage at the junction of vascular and avascular retina was also reported but was not commonly seen [1]. Interestingly patient with unilateral primary congenital glaucoma can have bilateral peripheral retina nonperfusion[1]. The relationship between abnormal peripheral retinal vasculature and childhood glaucoma is not clearly understood. Hanna *et al*. hypothesized that vascular abnormalities in children with glaucoma is due to the decrease in ocular perfusion pressure [1].

Blair *et al*. found that in children up to 13 years of age, the avascular retina can be a normal variant [2]. It can extend up to 1.5 disk diameters (DD) temporally and 1 disk diameter nasally from the ora serrata [2]. However, ≥2 DD of nonperfusion should be considered abnormal and a sign of peripheral nonperfusion. Hanna's studies demonstrated larger areas of retinal nonperfusion in patient with congenital glaucoma than those reported by Blair *et al*.

A study by Essam *et al*. found 13 patients had rhegmatogenous retinal detachment in the contralateral eye with primary congenital glaucoma [3]. However, there were no identified retinal holes, breaks, and/or tears in those patients. The relationship between these 2 conditions was unknown.

Vascular nonperfusion of the peripheral retina has previously described in Coats' disease [4]. Even though the association of congenital glaucoma and coats' disease is unclear however the similar findings of vascular nonperfusion of peripheral retina maybe related. The most common treatment modalities for Coats disease are laser photocoagulation and cryotherapy [5]. However these treatments have limited success in the presence of exudation [4]. Our patient had stage 3b when the diagnosis of Coats' disease established. Intravitreal anti vascular endothelial growth factor (VEGF) and orbital floor triamciniolone were given. In Coats disease the level of VEGF is raised [4,6]. Anti-VEGF agents are believed to work by stabilising the blood-retinal barrier hence reducing the amount of subretinal exudation. Intravitreal triamcinolone reduces subretinal fluid and exudates.

Therefore, it can facilitate the application of other therapies for vascular pathologies [4].

## CONCLUSION

Patient with unilateral congenital glaucoma may have bilateral retina vascular abnormality. This may lead to other retinal problem in future. Therefore, regular examination of the retina is mandatory in such patient.

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