Have You Ever Looked Inside A Child’s Eye? - A Pictorial Review from The Royal Liverpool Children Hospital, Alder Hey, Liverpool

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Learning objectives

The aim of this poster is to expose radiologists, particularly radiologists in training, to a range of benign and malignant, acute and non-acute paediatric intra-orbital disease manifestations. For each pathology, a brief description of the mode of the patient's presentation and the typical characteristics observed while performing the ultrasound examination will be described.
Background

Clinical examination of the paediatric population is at times very challenging for the clinicians. This is particularly pertinent to ophthalmic examination of a child who may find this examination uncomfortable and distressing. Prompt and accurate diagnosis of intra-orbital pathology is essential to avoid potentially devastating consequences and long term sequelae in such a vital organ.

The clinical radiologist has the unique capacity of being able to view the globe with the eyelid closed using a variety of imaging modalities. Due to its lack of ionising radiation and its high resolution, ultrasound is an ideal first-line tool for viewing the eye when the child is reluctant to allow direct visualisation by the ophthalmologist or when direct visualisation is impeded by the pathology in question.
Clinically, congenital cataracts can manifest in various ways; clinicians identifying a cloudy white pupil when screened in the first 24 hours of life or the parents noticing the child being unable to focus on objects or having to hold their head at a certain angle to be able to see objects. Parents may also report asymmetrical red eye on photographs, where the affected eye fails to produce a red reflex.

Congenital cataracts may be hereditary or have an infective aetiology and are frequently bilateral when present. Typically, sonographically, a well defined oval area of abnormal echogenicity and increased thickness is described in the central, posterior part of the lens; no associated vitreous or retrobulbar abnormalities are noted (Figures 1, 2). Surgical removal of the cataract is usually the treatment of choice.

A coloboma results from incomplete closure of the embryonic choroidal fissure which occurs at approximately the 6th week of gestational development. The key-hole shaped defect may affect the lens, iris, retina, choroid, macula or eyelid and affects both eyes in 50% of cases. Colobomas may present, clinically with poor vision, however in some cases vision may be entirely normal. Colobomas are sometimes discovered incidentally on CT/MRI by virtue of their association with other disease entities for which the patient is being investigated, such as an encephalocoele or corpus callosum agenesis. On ultrasound, an anechoic or hypoechoic outpouching is noted at the site of the optic disc (Figure 3). If the condition is unilateral, a discrepancy in size of the globes is noted, with the globe containing the coloboma being the smaller one. There is currently no treatment for colobomas; continued monitoring of growth and regular checks for associated complications of retinal detachment (as in our case) and glaucoma are however carried out.

Drusen, often a familial condition, may be an asymptomatic condition but patients can present with headaches and visual field impairment. It is caused by accumulation of hyaline material on or in the vicinity of the optic disc. In our case, bilaterally optic disc elevation- pseudopapilloedema - was noted on fundoscopy. An MRI was performed which did not reveal any optic disc pathology. On ultrasound however, echogenic foci with characteristic distal acoustic shadowing in keeping with calcification was identified at the optic discs bilaterally (Figure 4). No definite treatment is available for drusen, however regular screening for associated angiod streaks and retinitis pigmentosa should be carried out.

Persistent hyperplastic primary vitreous (PHPV) is a rare condition in which arrest of the normal regression pathway causes persistence and proliferation of embryonic hyaloid vascularity in the primary vitreous. Clinically, unilateral leukokoria (white pupil) is usually
evident and a serpiginous tubular mass extending from the lens posteriorly to the optic disc is noted on fundoscopy. However, the association with cataracts may impede the ophthalmologist from noting this characteristic clinical finding. On ultrasound, as in our case, an abnormally shaped and echogenic lens (cataract) together with a band of echogenic tissue containing small vessels as seen on colour doppler, extending from the posterior lens capsule to the optic disc is seen (Figure 5, 6). Surgical treatment is the treatment of choice.

Papilloedema (swelling of the optic disc), is one of the clinical clues to the presence of increased intracranial pressure. Prominent optic discs without the presence of calcification (hence excluding drusen), as seen on ultrasound can confirm the presence of this fundoscopic finding (Figures 7, 8).

Retinoblastoma is the most common intraocular malignant tumour in the paediatric population. Most cases of this rare neoplasm and nonheritable, however some are of the heritable sporadic variety and a few are familial (trilateral retinoblastoma: bilateral retinoblastoma and a pineoblastoma, quadrilateral retinoblastoma: trilateral retinoblastoma with an additional neuroectodermal malignant focus in the suprasellar cistern). Most cases present at 18 months of age with leuokoria and absence of the normal red reflex on examination, strabismus or proptosis most commonly. Characteristic ultrasonic features are of a mixed echogenicity, heterogenous, solid intraocular mass in the posterior globe. Hyperechoic areas with posterior acoustic shadowing are in keeping with calcific foci and anechoic appearances are likely to represent tumour necrosis (Figures 9, 10). In our case, the mass was noted to be fixed to the posterior wall of the globe and there was associated stranding in the vitreous (likely to represent the known association with vitreous haemorrhage). Distortion of the normal globe architecture was also identified (Figure 11). The finding of calcification has been found to be a favourable prognostic sign. Sonographic appearances of retinoblastoma were confirmed on subsequent CT and MRI imaging. Management is tailored for each individual patient, with chemotherapy, radiotherapy and surgical removal of the eyeball all being therapeutic options individually or in a combined fashion. Spontaneous regression has been described in approximately 1% of cases.

Retinal detachment secondary to trauma, an inflammatory process or tumour may be complete or partial (one intraorbital quadrant only). Typically patients report sudden onset of a veil-like opacity in their visual field. On ultrasound, multiple thick curvilinear echogenic septations are seen surrounding loculated pockets of fluid. The bands as in this case are typically non-mobile and tethered to the optic disc posteriorly and in complete retinal detachment assume a V-shaped configuration (Figures 12, 13, 14).
Fig. 0: Congenital cataract

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**Fig. 0:** Congenital Cataract

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**Fig. 0:** Coloboma

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**Fig. 0:** Drusen

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**Fig. 0**: Persistent Hyperplastic Primary Vitreous

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Fig. 0: Persistent Hyperplastic Primary Vitreous: Colour Doppler Imaging

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Fig. 0: Papilloedema

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**Fig. 0:** Papilloedema

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Fig. 0: Retinoblastoma

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**Fig. 0:** Retinal Detachment

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Conclusion

Paediatric eye pathology can be catastrophic if not diagnosed and managed promptly and correctly and the Radiologist plays an important role in the care of these children.

Cross sectional imaging is a well recognised imaging modality for imaging the eye, but there is less awareness that ultrasound plays an important role, particularly in the paediatric population.

Ultrasound can be an invaluable diagnostic tool and the aim of this review is to raise awareness of this imaging technique and to demonstrate the pathologies encountered.
References