Standardised Echography In The Diagnosis & Assessment Of Congenital Glaucoma

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Introduction

- Primary Congenital Glaucoma (PCG) is a rare condition affecting 8 in every 10,000 births.[1]
- There is a direct correlation between increasing IOP and increasing axial length, and therefore PCG is characterized by abnormal ocular growth during infancy.
- Studies have shown that the eye in PCG has a mean axial length some 25% longer than normal in infancy, and can increase to around 50 % longer with age.[1]
- Axial length measurements in PCG were first assessed in 1969 by Gernet and Hollwich.[2] Since then a direct correlation between axial length and the severity of visual field loss has been established.
- It is thought that increased IOP in childhood occurs as a result of an abnormal increase in axial length, an increase in AC depth, narrowing of the lens, and an increase in corneal diameter, [3] the cornea also becoming shallower.
- As the eye becomes larger, the sclera becomes thinner, and the effect of the increased IOP is to stretch and thin the neural rim, resulting in increased nerve fibre loss. Visual field loss usually occurs some 5 years later.[4]
- This study uses the latest echographic technology in Standardised A and B scanning techniques to evaluate the dimensions of the eye in PCG, but also to look at structural changes in the optic disc and optic nerve.

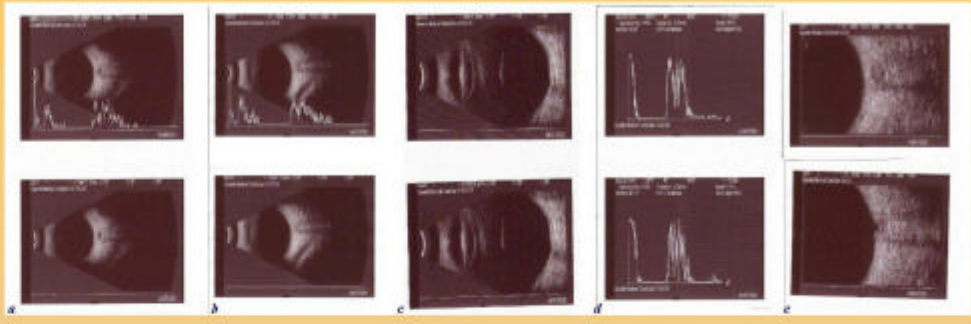

Fig a. Normal Optic Nerve Diameter & reflectivity. Fig b. Abnormal optic nerve diameter & reflectivity in PCG. Fig c. Measurement of AC depth & AL in a patient in PCG. Fig d. Inner Pial Reflectivity & diameter in same patient using standardised A-scan. Fig e. B scan of a patient with PCG showing a deeply cupped disc & reduced OND.

Methods

- Thirty two patients with PCG (16 females 16 males), mean age 13.7 years (age range 2 Months to 26 years) underwent Standardised A and B scan echography using a Quantel Cine Scan analogue long focal length scanner.
- This scanner has the advantage of giving high resolution images of the optic disc and proximal optic nerve, and because of its unique Time Gain Control features can give accurate imaging of the anterior chamber. This meant that accurate axial length measurements could be obtained in young children without the need to resort to anaesthesia, as measurements could be made through the closed eye lid.
- Measurements of optic nerve diameter (OND) and inner pial reflectivity (IPR) were made using the technique described by Ossoinig. [4] Essentially A or B Scan probes were placed at the lateral canthus of either eye and aimed towards the orbital apex. Scanning in a vertical transverse mode resulted in a perfect cross sectional image of the optic nerve approximately one cm behind the disc.
- The Anterior Chamber depth (AC), Axial length (AL), Optic disc cup depth (CD), optic nerve diameter, and inner pial reflectivity were measured in each eye, and compared to those values obtained from 20 age and sex matched controls (mean age 14.3 years - 10 males, 10 females).
- Group mean values were obtained for each paradigm, and compared to controls using Analysis of Variance.

Discussion

- As expected ocular dimensions were greatly increased in patients with PCG, AC depth and AL being greater than normal ranges in all patients.
- These findings confirm those of other authors in showing that increased ocular dimensions are a crucial factor in this condition.[1]
- In addition to this, the finding of a marked increase in cup depth, and a marked reduction in optic nerve diameter, combined with increased reflectivity of the inner pial portion of the optic nerve, was a clear indicator of significant nerve fibre loss and optic atrophy in these patients.
- Many of these patients had normal or near normal visual fields, despite the structural changes in the optic nerves, which confirms the findings of other authors that significant optic nerve damage due to high IOP in combination with scleral thinning resulting from increased ocular dimensions, can occur some time before visual field loss occurs.[5]
- This technique of identifying nerve fibre loss and atrophy as we have shown in previous studies [6] is of great value in infants or young children in whom visual field analysis may not be possible, and in any case the absence of field loss does not correlate with a healthy optic nerve.
- Sampaolesi [3] has shown that repeated axial length measurements in children with PCG is of great value in determining the best management of these patients and also as a prognostic indicator.
- As our results show there is a direct correlation between optic nerve damage and increased ocular dimensions, and therefore echography has an important role to play in the management of these patients.

Results

- Group mean AC depth, and AL were significantly greater than controls ($p<0.001$)
- All AL measurements being greater than the uppermost normal range for the age (see figure).
- 85% of AC depth measurements were greater than the uppermost normal range.
- Optic disc cup depths (CD) were significantly greater than controls ($p<0.001$) (see table).
- Group mean OND values were significantly ($p<0.001$) less than control values, and group mean IPR values were significantly higher than controls ($p<0.001$).

	PCG (n=64)	Control (n=40)
AC (mm)	4.9 (0.6) *	2.9 (0.25)
AL (mm)	26.8 (2.7)**	23.2 (1.8)
CD (mm)	1.50 (0.17)**	0.30 (0.05)
OND (mm)	1.74 (0.12)**	2.90 (0.25)
IPR (%)	65.7 (6.8)**	32.40 (3.8)

Table 1. To show group mean values of each parameter ($p<0.001$ compared to controls, ** $p<0.005$ compared to controls)*

References

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