

Giant Retinal Tears in Children: Associations and Outcomes

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Purpose: Giant Retinal Tear (GRT) as a cause of retinal detachment is uncommon in children. Their associations and surgical outcome in children of 16 years and less are not well described.

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Material and Methods: All patients who presented with retinal detachment due to GRT between 1998 and 2004 were included in the study. They underwent 3 ports Pars Plana Vitrectomy, 360 encirclement with 2.5 mm band and fluid perfluorocarbon liquid silicone oil exchange.

Results: 12 patients, 8 boys and 4 girls with an age range of 5-16 years presented and underwent surgery during this period. High myopia, Sticklers' syndrome, Weil Marchisani syndrome, trauma and Anterior Segment Cleavage disorder were the principal associations. In five patients the fellow eye was already NPL due to chronic retinal detachment. 7 patients had minimal or no PVR (Proliferative Vitreo-retinopathy) while the others ranged from stage B to C2 PVR. While all except one retina were re-attached per-operatively, at six months only 4 retinas remained fully attached. Both children under 10 years of age had a poor outcome.

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Conclusion: Retinal detachment, due to GRT, in children often has poor outcome due to aggressive evolution of PVR and difficulty in posturing. Younger age may be associated with poorer outcome.

Tears that extend 90° or more around the circumference of the retina are called giant retinal tears^{1,2}. Vitrectomy with internal tamponade is the treatment of choice for them. Due to wide exposure of retinal pigment epithelium (RPE), PVR³ is more common in retinal detachment due to GRT. In adults GRT is often associated with myopia and Sticklers syndrome among others. At least in adults with GRT in one eye, 12% incidence of giant tear formation in the fellow eye has been reported⁴. The incidence, associations and outcome of Giant tears of the retina in the population under 16 years of age are not well described as the frequency of rhegmatogenous retinal detachment in children is reported to be 1.7-5.9%⁵⁻⁷ of all retinal detachments.

Our aim is to study the associations and surgical outcome of retinal detachment in patients under 17 years of age.

MATERIAL AND METHODS

In a prospective hospital based interventional case series spanning seven years from 1998-2004 we studied all children who presented with a rhegmatogenous retinal detachment to the retinal unit of the Lahore General Hospital. Type of retinal tear, associated ocular and systemic features and outcome of surgery were recorded. The minimum follow-up was six months. The main outcome measure was anatomic retinal re-attachment.

RESULTS

In this five year period 83 children under 16 years of age were treated for retinal detachment. Of them 12 (14%) children (8 boys & 4 girls), had retinal detachment due to a GRT. Their age ranged from 5-16 years (Table 1).

The time from onset of retinal detachment to presentation ranged from 13 to 90 days and the distance patients traveled to seek treatment ranged from 0 to 200 miles. The significant associations (Table 2) were high myopia in three patients, Sticklers syndrome in three patients, Weil Marchisani syndrome, cataract surgery, trauma and anterior segment cleavage disorder in one patient each. No association was found in two patients.

Seven patients had minimal or no PVR (Proliferative Vitreo-retinopathy), one patient had stage B and 4 patients were in various levels of stage C (Table 3). While all except one retina was re-attached per-operatively, at six months only 4 retinas remained

fully attached. Both children under 10 years of age had a poor outcome.

The fellow eye was already no light perception in 5 patients (41%) due to chronic retinal detachment.

At the end of six months only four out of 12 retinas were anatomically fully attached. One patient underwent three re-operations for recurrent retinal detachment. Silicone Oil could be removed successfully in only one patient.

DISCUSSION

Posterior vitreous detachment (PVD) and abnormal vitreo-retinal adhesions are considered the principal underlying reasons for retinal detachment. The incidence of retinal detachment rises as the frequency of PVD increases⁸. In children as the vitreous is intact, so the frequency of retinal detachment is also low. Retinal detachment due to GRT is especially uncommon in children and therefore this problem is not well documented in the literature.

The eye department of the Lahore General Hospital is a tertiary referral center in Punjab, the most populous province (70 Million) of Pakistan. This center has special interest in vitreo-retinal surgery in an area with otherwise limited facilities for retinal problems. Due to a large referral base we were able to gather this case series.

We report a series of 12 patients who presented to us over a period of 7 years with retinal detachment due to GRT. A total of 83 patients less than 16 years of age were operated on during this period. In this series 14% of all paediatric retinal detachments were due to GRT. The time delay from onset of retinal detachment to presentation ranged from 13 to 90 days. This is explained in part by the distance the patients had to travel to seek treatment i.e. 0-200 miles. Poverty, limited access to facilities and perhaps late reporting by children^{9,10} were other significant factors.

Table 1: Age of Children presenting with GRT

Age	No. of Patients
5-6 years	1 patient
7-8 years	Nil
9-10 years	2 patients
11-12 years	1 patient
13-14 years	4 patients

15-16 years	4 patients
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Table 2: Associations of Giant Retinal Tears

Associations	No. of Patients
High Myopia	3 Patients
Sticklers Syndrome	3 Patients
Weil Marchisani Syndrome	2 Patients
Trauma	1 Patient
Anterior Segment Cleavage Syndrome	1 Patient
Cataract Surgery	1 Patient
No Association	1 Patient

Table 3: Stage of PVR at presentation

Stage of PVR	No. of Patients
No or Stage A PVR	7 patients
Stage B PVR	1 Patient
Stage C PVR	4 Patients

10 out of 12 patients had a significant ocular or systemic association, the most common being myopia and Sticklers syndrome Table 1. Two patients had no clear cut association. Of great concern were the findings that, 5 out of 11 patients had their fellow eye already affected by retinal detachment and had no light perception. Two out of three patients with Sticklers and the only one with Weil Marchisani belonged in this group.

The delay in presentation and the nature of the disease meant that 4 out of 12 had stage C PVR on presentation. A reflection of the aggressive nature of the disease is witnessed in the anatomical surgical outcome. While per-operative retinal re-attachment was achieved in 11 out of 12 cases, at the end of six months only 4 retinas (33%) were still fully attached. Silicone oil could be removed successfully in only one patient. One of these patients has had three re-operations involving membrane peel and retinectomy and cataract extraction to preserve retinal attachment. Both patients under 10 years of age re-detached irreversibly.

Previous studies evaluating the surgical outcome after GRT show variable success rate. Scott IU et al¹¹ achieved an anatomical success rate of 68 % in their case series of 34 children under 16 years of age with GRT and preservation of ambulatory vision in 53 % of these children following vitrectomy and internal tamponade with silicon oil at 6 months. In a similar aged population Karel I et al¹² had a success rate of 82 % at 2 years which declined to 45 % at 5 years in 24 eyes of 22 children.

The principal cause of failure in our study was the aggressive PVR¹³. Inability to induce PVD anterior to the equator would perhaps be another cause. Residual vitreous may have provided the scaffold, which resulted in shortening antero-posteriorly as well as circumferentially. Inability to maintain posture for internal tamponade by silicone oil probably also played an important part¹⁴.

In conclusion GRT in the paediatric age group though uncommon have a significant chance of affecting both eyes and has a poor prognosis in terms of anatomical outcome in the medium term.

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