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The aetiology of paediatric rhegmatogenous retinal detachment: 15 years experience

Abstract

Purpose To provide contemporary data on the aetiology of paediatric rhegmatogenous retinal detachment (RRD) in the UK population. Methods Retrospective case series. Results Eighty-eight eyes in 82 patients (aged 0-16 years) were treated for RRD at Bristol Eye Hospital between 1 January 1990 and 31 December 2004. Seventy-three per cent of patients were male and the main predisposing factors were trauma (53%), associated conditions (27%), and high myopia (17%). Nineteen per cent of RRDs were idiopathic, and the majority of these were due to inferotemporal dialyses. The macula was detached on presentation in 66% of eyes. Conclusions The principal causes of paediatric RRDs have not changed over the past 40 years. Those due to congenital cataracts, retinopathy of prematurity, uveitis, and glaucoma are now less prevalent, presumably reflecting advances in their management. Differences with other contemporary series may arise from geographical variation in the prevalence of myopia and other associated conditions, as well as institutional referral patterns. Full examination of the retinal periphery is advised for children with eye injuries (to exclude dialyses). Eye (2008) 22, 636-640; doi:10.1038/sj.eye.6702724; published online 9 February 2007

Keywords: child; adolescent; eye injuries; degenerative myopia; retinal detachment; risk factors

Introduction

Rhegmatogenous retinal detachment (RRD) is uncommon in children and its aetiology differs

from retinal detachment in adults.¹⁻⁷ In the 1960s and 1970s (when the largest case series of paediatric RRDs were published) trauma, myopia, aphakia, and retinopathy of prematurity (ROP) were the leading causes.^{8–12} We would expect this to have changed with subsequent advances in the management of congenital cataract13 and ROP,14 and the reduced incidence of ocular trauma following the introduction of seat belt legislation.^{15,16} Recent reports have either examined small patient numbers,^{17–20} focused on particular aetiologies,^{21–23} specifically considered the management of complex cases,^{24,25} or reviewed populations in the Far East,^{26,27} where myopia (even in the paediatric age group) is reaching almost epidemic levels.²⁸ Contemporary data are needed to develop strategies for prevention and early detection of this sight-threatening condition in European populations. This paper provides this, reporting a large series of paediatric RRDs from a single tertiary referral centre in the UK.

Patients and methods

Consecutive patients with RRD aged 0-16 years presenting between 1 January 1990 and 31 December 2004 to Bristol Eye Hospital (a regional tertiary referral centre for Southwest England and South Wales, covering a population of approximately 5 million people) were identified from a manual review of theatre log books. All records were available for this 15-year period, and patients were identified by their date of birth, procedure, and surgeon. If the diagnosis was unclear (including 'examinations under anaesthesia'), the patient's clinical notes were reviewed. A standard data set was then recorded for each patient. This included age at presentation, sex, affected eye, history of trauma, refractive status, family

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history, type of detachment and retinal break, macular status (on or off), and coexisting ocular or systemic diagnoses. Only the initial presentation was included for eyes which had more than one procedure.

Patients with non-rhegmatogenous causes of retinal detachment (including Coats' disease, ocular tumours, and inoperable traction retinal detachments secondary to ROP) were excluded from this study.

Results

Patient characteristics

Eighty-eight eyes in 82 patients (age 0–16 years) were treated for RRD at Bristol Eye Hospital between 1 January 1990 and 31 December 2004. Sixty (73%) patients were male and 22 were female. Their median age was 14 years (Figure 1). Forty-one (47%) right eyes and 47 left eyes were affected.

Trauma

Forty-seven (53%) cases were associated with trauma (Figure 2), including penetrating injury in 10 (11%) eyes. Thirty-seven (79%) of these traumatic retinal detachments occurred in males. The time between injury and diagnosis ranged from less than 24 h to 3 years.

Other associated conditions

In total, 24 (27%) eyes were associated with ocular or systemic comorbidity, and of the five patients in this group who had retinal detachment surgery in both eyes, one was bilaterally pseudophakic (following congenital cataracts), two had Stickler's syndrome, one had atopic dermatitis, and one had cerebral palsy. Of the unilateral cases, two eyes had uveal colobomata, one in association with Wildervanck syndrome and the other in association with contralateral microphthalmos. One case had Wagner's syndrome, one had Marfan's syndrome, and three had X-linked juvenile retinoschisis. Three other cases were associated with non-familial abnormalities (microcephaly with cataract, corectopia with lens



Figure 1 No separate title required (this is incorporated in the figure).

dislocation, and retinal necrosis following herpes simplex retinitis). In the absence of 'associated conditions' or myopia, there was never a family history of retinal detachment.

Myopia

Twenty-five (28%) eyes were myopic. Fifteen (17%) were high-myopes with a refractive error of $\ge 6D$ (range -6 to -22D). Five of these highly myopic eyes were in three patients with Stickler's syndrome. Two highly myopic eyes had associated ROP.

Four of the 25 retinal detachments in myopic eyes were also associated with trauma.

'Idiopathic'

Seventeen (19%) eyes were neither associated with trauma, high myopia, nor any identified associated condition. Thirteen (76%) of these eyes had dialyses.

Retinal features

The frequency of different types of retinal break is illustrated in Figure 3. Of the 38 dialyses, 24 (63%) had a clear history of previous ocular injury, as did six (67%) of the nine giant tears. Multiple holes and tears were characteristic of myopic RRDs. The macula was detached in 58 (66%) eyes.



10% High myopia & associated conditions

Figure 2 Aetiology of paediatric RRD.



Figure 3 Types of retinal break.

The clinical records of all patients identified from the theatre log were successfully retrieved and reviewed. There were no missing data with respect to high myopia. In patients without a history of amblyopia refractive error was assumed to be equivalent to the contralateral eye. The presence or absence of retinal breaks was not recorded in 2% of cases (Figure 3).

Discussion

The principal similarities between this and older reports^{8–12} are that (i) trauma is still the leading cause of paediatric RRD; (ii) there is a male predominance; (iii) chronic detachments and late presentation remain a problem; (iv) retinal dialyses account for most idiopathic presentations; and (v) the frequency of RRD increases with age.

Trauma accounted for 44% of Winslow and Tasman's series of 187 paediatric RRDs over 8 years,⁹ 42% of Hilton and Norton's report of 71 cases over 12 years,¹¹ 65% of Chen and Dumas's report of 181 cases over 10 years,¹⁰ and 34% of Daniel and Kanski's report of 163 cases over 15 years.⁸ Our proportion of paediatric RRDs attributable to trauma remains remarkably similar.

The prevalence of boys has always been associated with their predisposition to trauma because they are more likely to engage in dangerous activities than girls, and we would not expect this behavioral risk to have changed over the past 40 years. Consistent with this, 84% of all retinal detachments occurred in boys in Winslow and Tasman's series,⁹ compared with 79% of ours. This sex discrepancy persists in the non-traumatic group of detachments (26 male *vs* 15 female in our series), which is in-keeping with the findings of others;²⁷ however, the reason for this remains unclear. One possibility is that many of these 'non-traumatic' detachments are in fact due to previous trivial or forgotten injury.

Hilton and Norton,¹¹ and others,^{8–10,12} noted the high proportion of children presenting with chronic retinal detachments, a problem which remains today. Time from injury to presentation was up to 3 years in this series, and the macula was detached in 66% of cases (compared with 41% in reports of RD in all age groups¹).

One-third of dialyses were not associated with trauma, and 76% of the idiopathic retinal detachments were due to dialyses. Previous series reported similar findings,^{8,9,11} suggesting that 'spontaneous' dialyses are a separate clinico-pathological entity in the paediatric age group. These characteristically occur in the temporal retina, which is developmentally the last area to vascularise, and consequently it has been suggested that they may occur in children with a congenital predisposition.²⁹ However, it is difficult to prove that these dialyses are not the result of previous trivial or forgotten trauma and the true underlying pathology remains unproven.³⁰

The incidence of RRD from all causes is known to increase with age¹⁻⁷ and paediatric RRD series are inevitably biased toward older children with 'adult-type' pathology.³¹ It is important to consider this when comparing published data because the age range used to define 'paediatric' varies from 0-14 to 0-20. Reports before 1970 found two peaks in incidence; below 6 years of age due to 'congenital' causes, and above 6 years due to trauma.³ The very small number of children under 6 years of age in our series probably reflects advances in the management of congenital cataracts, ROP, and premature neonates. None of our children was aphakic and one was pseudophakic (with RDs in both eyes emphasising an additional predisposition in this case). Only 2% or our series had ROP, compared with 8% of earlier RRD series.9 Furthermore, we had no cases of paediatric glaucoma, just one with uveitis, and few bilateral RRDs in myopes; possibly because all fellow eyes received prophylactic treatment at the time of surgery.32

Small contemporary reports from the UK²⁰ and Israel¹⁹ corroborate our findings with a similar range and distribution of aetiologies: 44 and 56% traumatic, 20 and 31% myopic, respectively. In the UK group,²⁰ 80% were due to trauma, myopia or associated comorbidity, and the remaining 20% had inferior dialyses; 47% were found incidentally or presented late.²⁰ Eighty per cent of the Israeli series were male and 81% presented with macular detachment.¹⁹

638

In contrast with our findings, uveitis, ROP, and buphthalmos accounted for 15, 12, and 6%, respectively, of a recent report of 39 eyes over 10 years.¹⁸ High frequencies of these pathologies were also found in another small American series,²² and in a recent report from Japan (of 49 patients over 14 years), familial exudative retinopathy and atopic dermatitis accounted for 13 and 9% of cases.¹⁷ Myopia was also the leading cause of paediatric RRD in two large series from Taiwan published last year.^{26,27} These disparities are attributable to local differences in the prevalence of myopia²⁸ and referral patterns, illustrating the limitations of retrospective studies such as this from individual centres.

It is important to emphasise that the data presented should not be considered to be representative of all paediatric RDs, as non-rhegmatogenous causes were excluded. Hence, although the incidence of ROPassociated RRD appears to be falling, this may not be borne out in an analysis of all paediatric cases (which would also include non-rhegmatogenous traction RDs). Furthermore, this series only pertains to RRDs, which were treated or assessed in the operating theatre, as cases judged to be inoperable without examination under anaesthesia would not have been identified from the surgical log. However, as this is exceptionally rare, we consider the risk of significant bias from incomplete data acquisition to be minimal.

In summary, this report from a single UK tertiary referral centre suggests that the primary causes of paediatric RRD (trauma, associated conditions, and myopia) have not changed over the past 40 years, although the contribution of congenital cataract, ROP, uveitis, and glaucoma has reduced, presumably reflecting advances in the management of these conditions. Differences between this and other contemporary series are probably explained by geographical variation in the prevalence of associated conditions and myopia, as well as variation in institutional referral patterns. An analysis of visual outcomes after surgical repair has not been presented as the heterogeneity of aetiologies, severity of globe injuries, and evolution of surgical techniques over the study period confound the interpretation of this data.

Despite our knowledge that trauma and myopia are principal risks for paediatric RRD, there is no obvious solution to the problems of delayed presentation or the predisposition of boys to ocular injury, and we also have no data from which to justify routine screening of highly myopic children. We would, however, recommend that care is taken to perform a thorough examination of the retinal periphery when examining children with eye injuries; in particular to look for dialyses in the inferotemporal quadrant.

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