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INCIDENCE, RISK FACTORS AND OUTCOMES OF RHEGMATOGENOUS RETINAL DETACHMENTS REPAIR IN MARFAN SYNDROME

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Marfan syndrome is an inherited diseases which is commonly autosomal dominant and associated with abnormal variants in fibrillin-1 (FBN1) gene or less commonly in tumor growth factor-beta receptor 2 (TGFBR2). FBN1, which is coded on chromosome 15 in the q21.1 locus is an integral component of the extracellular myofibrils. 1,2
Ocular manifestations of Marfan syndrome include malpositioning of the crystalline lens (ectopia lentis), rhegmatogenous retinal detachment (RRD), glaucoma, myopia, and corneal abnormalities. However, ectopia lentis and RRD are common in Marfan syndrome patients and often require surgical management. 3,4
The prevalence of retinal detachment in Marfan syndrome varies across studies, with reported rates ranging from 4% to 54%. 4-6 Abnormal lens position, progressive myopia and vitreous liquefaction play roles in the predisposition of Marfan syndrome patients to RRD. 2,6
Surgical treatment outcomes for RRD in Marfan syndrome patients are variable and might be affected by several factors. It was suggested that the outcomes of RRD repairs are anatomically and functionally favorable. 4 However, with the advances in pediatric lens removal surgeries and secondary intraocular lens implantations, the pathogenesis and results of RRD repair might be influenced by the lens status. 7
In this study we aim to analyze the incidence, risk factors and outcome of retinal detachment in Marfan's patient from several perspectives and to determine the long-term anatomical and visual outcomes, taking into consideration the different factors which might play a role in the determination of outcomes.

Retrospective chart review including lens status, previous surgeries, details of intraoperative retinal findings and follow up outcomes. (mean ±SD) years.

163 eyes of 82 patients with Marfan syndrome were included. Among 82 patients with Marfan syndrome, 43 (52.4%) were males and 39 (47.9%) were females. The average age of patients was 30.55 ±14.6 years. The average duration of follow ups was 15.3 ± 13.4 years. 54 eyes (33.1%) of 45 patients had RRD. Bilateral RRD was found in 9 patients (20%). The average age at the development of RRD was 25.2 ±12.5 years. There was significantly higher risk of RRD among patients who had prior trauma (P=0.014), Previous ocular surgery (P=0.001), lensectomy without implantation of an intraocular lens (P=0.002) aphakia (P<0.001), lens subluxation (P=0.002) and higher Axial Length (P<0.001). For the 25 eyes which had prior ocular surgeries, the average duration between the first surgery and RD was 8.4 ± 6.9 years. Successful reattachment was achieved in 36 eyes (69.2%) while 16 eyes (30.8%) had recurrence of retinal detachment after 37.5 ± 41.1 months (3 years) and were successfully reattached with second repairs. There was a statistically significant improvement in final VA of eyes with RRD (1.3 ±1.0 snellen = 20\400), (P 0.001).

Eyes with Marfan Syndrome have a 33.1% risk of developing RRD upon long-term follow ups. Prior trauma, intraocular surgeries, aphakic status and high axial length are associated with higher risk. Anatomical outcomes are favorable in eyes without PVR.

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