Surgical interventions for the primary prophylaxis of retinal detachment in Stickler Syndrome: a systematic review

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INTRODUCTION
Stickler syndrome affects approximately 1 in 10,000 individuals. It is characterized by joint pain, facial abnormalities (eg. cleft lip and palate) and ocular abnormalities (eg. myopia). (Stickler 2001, 1965; Sneed 1999). As many as 60% of individuals with Stickler syndrome experience retinal detachment (RD) and, consequently, are at a high risk of blindness. While RD can occur at any age, and the risk is lifelong, the first RD in individuals with Stickler syndrome has been reported to occur most commonly in adolescence or early adulthood, between the ages of 10 and 30 years.

METHODS
A systematic review to assess the effectiveness of surgical interventions to prevent RD in children and adults with Stickler syndrome. A systematic search was made in October 2009 of 12 databases of published and unpublished literature: MEDLINE, MEDLINE in process; EMBASE; CINAHL; The Cochrane Library; Biological Abstracts; Science Citation Index; UK Clinical Trials Research Network (UKCRN); National Research Register archive (NRR); Current Controlled Trials; Clinical Trials.gov; Annual Meeting of the Association for Research in Vision and Ophthalmology. There was no restriction by language, date or study design (other than the requirement that studies have a comparator group).

Both studies performed prophylaxis in individuals either with no previous RD in either eye, or performed prophylaxis in the fellow eye of those with a previous RD in the primary eye. Each study reported a statistically significant difference in the rate of RD per eye between the groups receiving prophylaxis and the controls. Relative risks (RR) were calculated by the review authors based on event data reported (See Table 1). There was a statistically significant reduction in the risk of RD for those exposed to cryotherapy for bilateral prophylaxis compared to the controls (RR: 0.05, 95% CI 0.02, 0.14, p<0.0001), as well as for unilateral prophylaxis (RR: 0.16, 95% CI 0.05, 0.47, p=0.0009). There was also a reduction in the risk of RD for those exposed to laser treatment for bilateral prophylaxis compared to the controls (RR: 0.28, 95% CI 0.04, 1.84, p=0.19), as well as for those exposed to unilateral prophylaxis (RR: 0.13, 95% CI 0.01, 1.90, p=0.45), but the relative reduction in risk was not statistically significant in either case. Neither study reported any major or long-term adverse events or complications associated with the interventions.

RESULTS
The reduction in the risk of retinal detachment, based on the published data associated with the evaluated treatments, is large. However, both included studies have a high risk of bias. Future trials appropriate for rare conditions, such as non-randomized sequential allocation trials, are needed to reduce the lack of certainty surrounding the reported estimates of effect.

CONCLUSIONS
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REFERENCE