

Prophylactic Retinal Surgery for Children with Type I Stickler Syndrome



FORUM DISCUSSION

Stickler Syndrome Conference
23 Oct 2016

Our story

- 4 known cases within our family; father, brother, myself, my son
- All adults have experienced retinal detachments
- Only my father had a GRT and is blind in one eye
- Harvey is 4 years old and at risk of retinal detachments
- We have seen in clinic 3 surgeons:
 - Mr Stephen Charles - Manchester Royal Eye Hospital
 - Mr Snead - Addenbrooks
 - Mr Eric Ezra – Moorfields





Retinal Detachments in Type I Sticklers

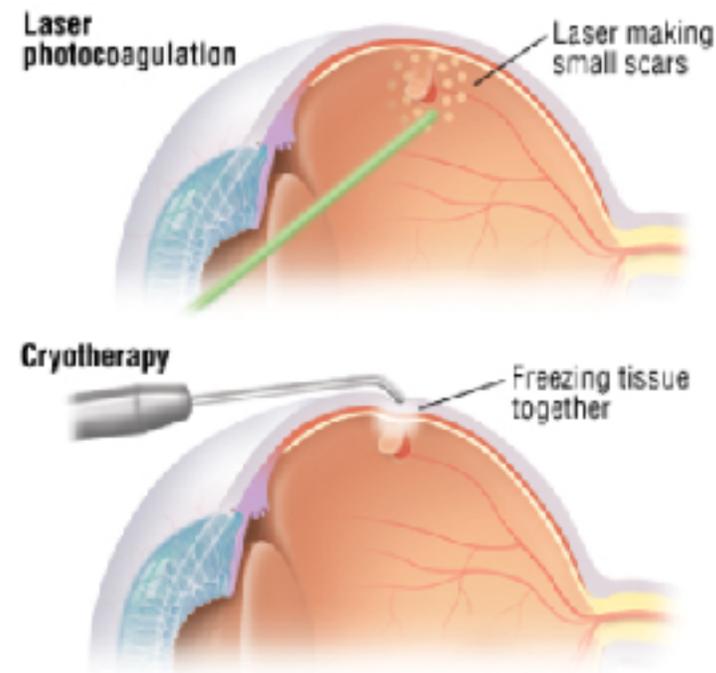
- Median age to first detachment maybe as young as 18 years
- High risk of retinal detachment
- Risk of Giant Retinal Tears before teenage years?
- Main risk in children is late presentation and resulting inoperability
- Loss of sight at an early age can lead to learning difficulties

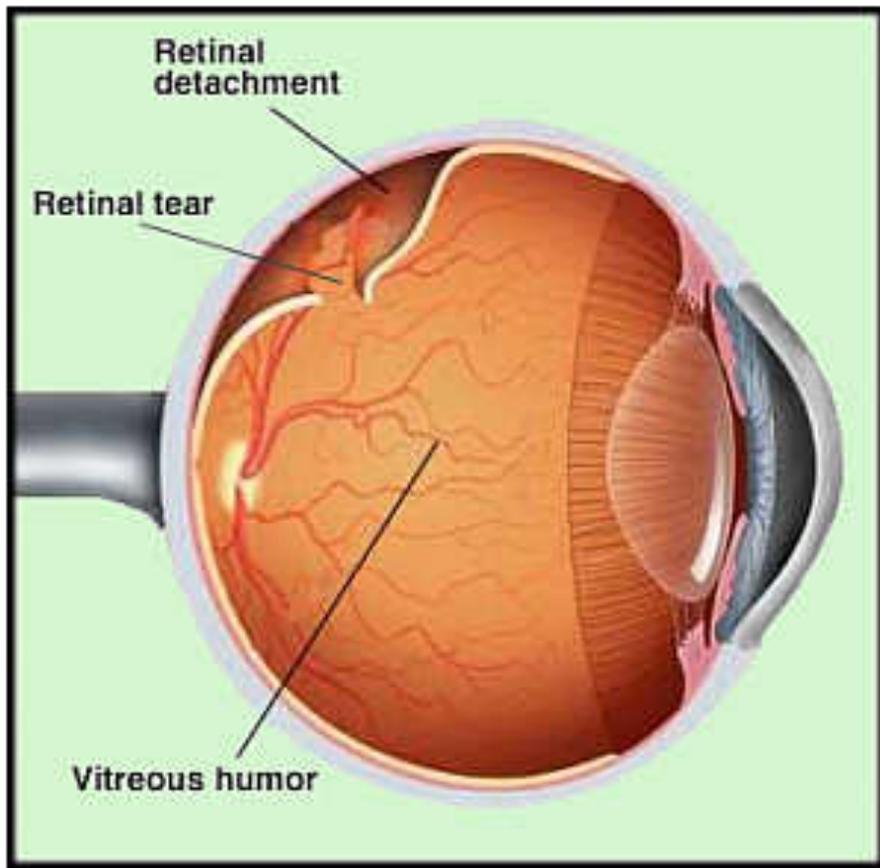
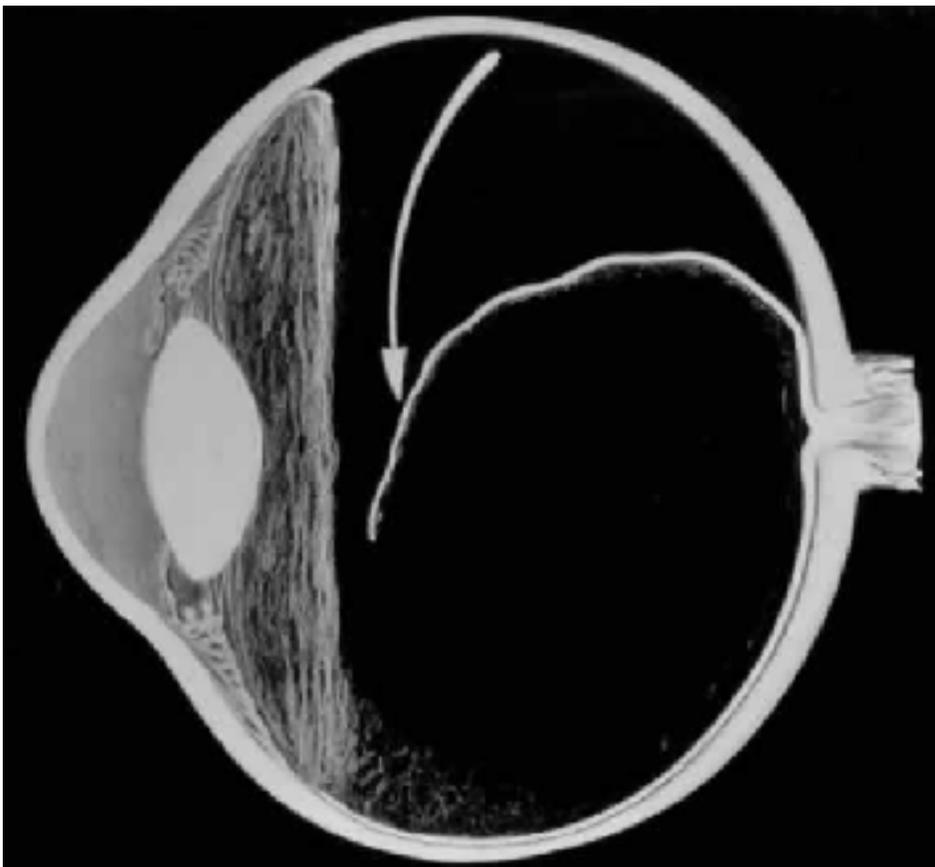




Prophylactic Surgery – What is it?

- 360° Cryotherapy or laser
- General anaesthesia
- Cryotherapy is performed by freezing the tissue from the outside of the eye
- Laser therapy is performed by directing a laser through a dilated pupil
- Both result in scarring of the retina at its edge
- Aim is to ‘stick’ the retina down to prevent a Giant Retinal Tear (GRT)





- The surgery will **not** prevent any retinal tears in the posterior (back) section of the retina, but only those originating from the anterior region (front)



Items for discussion

- What advice have you been given?
- What age to perform the surgery?
- What method to use? Laser, Cryo?
- What are your experiences of prophylactic surgery?
 - Age?
 - Side effects?
 - Issues?
 - Positives?
 - Overall confidence with the decision



Back up slides





Adverse Events (Side effects)

Types:

- Changes in visual acuity (clarity of vision)
- Lid and conjunctival inflammation
- Accommodation insufficiency (a reduced level of focus stamina required for accurate near binocular vision)
- Discomfort
- Photophobia
- Macular pucker (a layer of scar tissue that grows on the surface of the retina, particularly the macula, which is the part of your eye responsible for detailed, central vision). A *macular pucker* can cause blurred and distorted central vision
- Itchy eyes

Outcome

- Resolved?
- Time to resolution?

Mr Snead's results



Bilateral Matched

- 165 patients
- 9.1% vs 37% in control had detachments
- Mean age of bilateral matched groups at last review were 19.8 with SD of 15 (95% of people were between ages of 4.8 and 34.8)
 - Quite young mean age and chances of detachments increase with age

Unilateral Matched

- 39 patients (small group)
- 15.4% vs 69.2% in control had detachments
- Mean age of unilateral matched groups at last review were 31.4 with SD of 16.3 (95% of people were between ages of 15.1 and 47.7)



Mr Snead's results (cont.)

- Failure of prophylaxis occurred in 9% of patients (27/299)
- Average age of failure was 21.5 years
- Average time from treatment to failure was 5.6 years
- Of the failures
 - 4 were a break at the edge of the treatment (1 required further cryo, rest just laser)
 - 5 were anterior breaks going through the treatment barrier (requiring further cryo)
 - Rest were posterior tears
- Side effects may be under-reported due to nature of retrospective study
- Prevalence of retinal detachment in the control group was lower than published estimated from support group surveys



Questions

- Does the success rate increase or decrease compared with the age of prophylaxis? i.e. is there an increased risk of failure performing the surgery at a younger age?
- Risk of Giant Retinal Tears prior to teenage years?