

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

### Comment

Contralateral ARN is well described in cases with Herpes Zoster Ophthalmicus. Second eye involvement occurs in approximately a third of patients, typically within 6 weeks,<sup>8</sup> although fellow eye involvement decades following an initial infection has been described.<sup>9</sup>

Frances-Munoz *et al*<sup>3</sup> report an association between Kyrieleis' plaques and inflammation. In our case, the plaques increased as the vitritis and retinitis resolved. Interestingly, despite the high doses of steroids resolved. In our case, the Kyrieleis' plaques appeared to increase in number and spread around the optic disc while the overall inflammation was settling down. Eventually the plaques started to dissolve 4 weeks after presentation.

This case highlights the already known importance of examining the retina of the fellow eye when patients present with Herpes Zoster Ophthalmicus. In addition, the case provides further evidence that Kyrieleis plaques can potentially increase in size and number as inflammation settles down. The high-dose steroid treatment did not appear to have a role in limiting or reversing the development of these plaques.

### Conflict of interest

The authors declare no conflict of interest.

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Sir,

### Does the timing of treatment affect the ocular phenotype in patients with Mucopolysaccharidosis I homozygous for the L490P mutation?

Mucopolysaccharidosis I (MPS I) has autosomal recessive inheritance and has been associated in some cases with a single base mutation L490P.<sup>1</sup> Defective breakdown of glycosaminoglycans (GAGs) in MPS I causes accumulation in musculoskeletal, cardiorespiratory, and ophthalmic systems. In eyes, GAG deposition results in: (i) glaucoma, (ii) retinal degeneration, (iii) corneal haze, and (iv) optic disc swelling or atrophy.<sup>2</sup> We report 13 children (Table 1) homozygous for L490P, treated with either haematopoietic stem cell transplantation (HSCT) or enzyme replacement therapy (ERT). This is the first report to correlate ocular phenotype and timing of treatment in a subset of MPS I patients with a defined mutation.

### Comment

The effects of early treatment on the ocular phenotype in MPS are at present unclear. An animal study has demonstrated improved corneal clarity with ERT, particularly when started shortly after birth.<sup>3</sup> Previous retrospective case series of MPS I patients without defined genotype treated with ERT have not shown significant improvement in vision, corneal, or optic nerve changes.<sup>4,5</sup>

Little is known about genotype-ocular phenotype correlation in MPS I, but it may be expected that a series of patients with a single L490P mutation would have less ocular phenotypic variability than a group of patients with a variety of mutations. This case series demonstrates that the severity of corneal haze at final follow-up correlated with the timing of treatment with either ERT or HSCT (Figure 1a), although children treated earlier were younger at follow-up (Table 1). Timing of treatment did not correlate with retinal or optic nerve findings.

Children treated earlier had better final visual outcomes, although again they were younger at follow-up (Figure 1b). Three patients who were treated at younger than 60 months (cases 2, 5, and 8) showed improvement in vision, whereas none treated over 60 months of age had any improvement in vision and three had a deterioration (cases 9, 12, and 13). These results suggest that earlier treatment with ERT/HSCT for patients with MPSI may result in improved visual outcomes, but methods for objective assessment of ocular phenotype and longer follow-up of treated patients is needed to clarify this.

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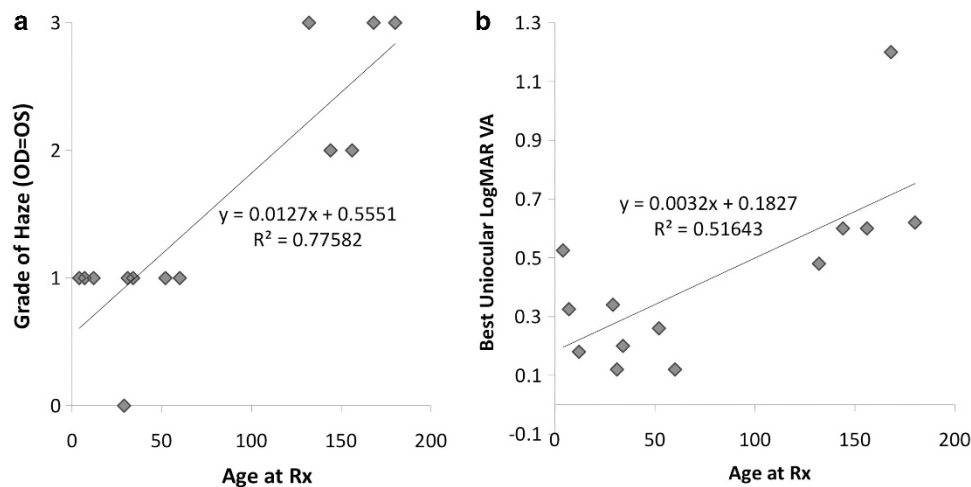
**Table 1** Clinical characteristics of patients homozygous for the L490P mutation

Case	Age at Rx (months)	Age 1st seen (months)	1st VA (OD, OS) (LogMar equivalent)	Corneal haze	Retina (OD, OS) <sup>a</sup>	Optic nerve (OD, OS)	Final VA (LogMar equivalent)	Age at final VA (months)
1	4	24	0.12 OU	Mild	N, N	N	0.55 OD, 0.65 OS	84
2	7	6	1.0 OU	Mild	N, N	N	0.33 OD, 0.38 OS	66
3	12	72	0.18, 0.47	Mild	N, N	N	0.18 OD, 0.48 OS	124
4	29	48	0.4 OD, 0.3 OS	Clear	N, N	N	0.34 OD, 0.44 OS	54
5	31	14	0.6 OU	Mild	N, N	N	0.12 OD, 0.37 OS	86
6	34	31	0.65 OD, 0.2 OS	Mild	N, N	N	0.65 OD, 0.2 OS	55
7	52	45	0.26 OD, 0.28 OS	Mild	N, N	Pale	0.26 OD, 0.28 OS	81
8	60	48	0.47 OU	Mild	N, N	N	0.12 OD, 0.3 OS	60
9	132	132	0.4 OU	Severe	N, N	N	0.6 OD, 0.48 OS	156
10	144	144	0.8 OD, 0.6 OS	Moderate	CR	PPA	0.8 OD, 0.6 OS	144
11	156	168	0.6 OD, 0.8 OS	Moderate	CR atrophy	N	0.6 OD, 0.8 OS	168
12	168	192	0.8 OU	Severe	N, N	<sup>b</sup> , pale	1.2 OD, 1.3 OS	196
13	180	180	0.47 OU	Severe	CR atrophy	N	0.62 OD, 0.8 OS	252

Abbreviations: CR atrophy, chorioretinal atrophy; N, normal; PPA peripapillary atrophy; Rx, treatment; VA, visual acuity (LogMar equivalent as measured with Cardiff cards, Kays pictures or ETDRS chart).

<sup>a</sup>Subjective grading of corneal haze.

<sup>b</sup>Not done.

**Figure 1** (a and b) Earlier age at treatment with ERT leads to reduced corneal haze and better final VA.

### Conflict of interest

JLA has received travel expenses and an educational grant from Biomarin Europe Ltd.

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Sir,

### What makes a good operation great? Factors determining patient satisfaction with local anaesthesia in cataract surgery

A Sub-tenon block (STB) is the most preferred choice of anaesthesia in the United Kingdom for cataract surgery. STB offers good akinesia and anaesthesia with a relatively low complication rate.<sup>1,2</sup> Nonetheless, STB has potentially serious complications including severe chemosis, and increased ocular pressure.<sup>3</sup> Since the late 90s, topical anaesthesia (TA) has been described as a safe alternative for injection anaesthesia in cataract surgery.<sup>4,5</sup>

We studied patient satisfaction with topical and sub-tenon anaesthesia and the factors that may affect patient satisfaction in cataract surgery.

A written questionnaire was distributed to 56 patients within 30 min post-operatively. The modified Iowa Satisfaction with Anaesthesia Scale (ISAS) was used, where patients responded to five statements (eg, 'I would want to have the same anaesthetic again') by placing a mark along a six choice vertical response column (eg, 'Disagree moderately'). In each statement, totally satisfied patient would score +3, a totally dissatisfied patient would score -3. The maximum and minimum score was 15 and -15, respectively. Additionally, patients were asked to score their level of pain on a Visual Analogue Scale (VAS) from 0–10, 0 being no pain and 10 being the worst pain experienced.

Analysis of the ISAS responses showed a mean score of 12.4 for the STB group (SD = 3.75) and 11.1 for the TA group (SD = 5.93), suggesting higher but not statistically significant ( $P = 0.399$ ) patient satisfaction in the STB

group. However, significant differences were seen in patient satisfaction with reduced duration of the operation ( $P < 0.002$ ) and seniority of operating surgeon ( $P < 0.004$ ). Post-operative pain evaluation using VAS showed no statistical difference between STB and TA groups. There were two cases (3.4%) of excessive pain experienced in TA group, requiring conversion to injection anaesthesia. In STB group, one patient had severe chemosis, one patient with minimal akinesia interfering with the surgery, resulting in reinforcement of anaesthesia.

We have found that there is no significant difference in patient satisfaction between the two groups. Patient satisfaction is, however, affected by the duration of the operation and seniority of the surgeon, which suggests that more complicated cases are best performed by senior surgeons.

### Conflict of interest

The authors declare no conflict of interest.

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