Eye (2011) 25, 818–827

www.nature.com/eye

CORRESPONDENCE

Sir,

Screening for symptomatic optic pathway glioma in children with neurofibromatosis type 1

We would like to clarify some important issues raised in the debate about screening children with NF1-associated optic pathway glioma (OPG).[1,2] The principles of population mass screening are not applicable to the detection of OPG in young children with NF1—a significant complication in a high-risk group of patients. Ophthalmological assessments aim to detect visual impairments resulting from a symptomatic OPG rather than to identify all NF1-associated OPG, at least half of which will never cause signs or symptoms.[3] Age-appropriate visual testing is recommended throughout the first decade of life to identify children requiring increased surveillance.[3]

MRI does not predict clinical behaviour or preclude the later development of an OPG; moreover, incidentally identified NF1-associated OPGs rarely progress or require treatment.[4] In the absence of refractive error, reduced visual acuity or proptosis in a child with NF1 is highly predictive of an OPG, while cataracts and retinal disease are rarely detected in this population. The significant causes of non-correctible visual loss in NF1 children are OPG, glaucoma secondary to a plexiform neurofibroma involving the eyelid, or proptosis and optic nerve damage due to a retro-orbital plexiform neurofibroma. While the latter two may be visible, detailed ophthalmological examination including fundoscopy is necessary to detect a symptomatic OPG.

In current practice, the goal of annual age-appropriate visual assessments in children with NF1 is to identify abnormalities in visual function attributable to an OPG. These assessments should continue until at least 7 years of age, when the risk of visual impairments from OPG is significantly reduced.[4,5] The finding of non-refractive decreased visual acuity warrants neuroimaging for identification and localisation of OPG, followed by serial visual testing and neuroimaging. Visual progression, defined as a two-line decrement in visual acuity, is an indication for chemotherapy.

Recently, the National Commissioning Group funded a 'Complex NF1' service in London and Manchester. A cardinal aim is to work with local ophthalmologists and optometrists to perform annual visual screening for OPG in NF1 children under 8 years. This prospective assessment will facilitate a nationally cohesive screening programme for OPG in a high-risk group of children.

Conflict of interest

All the authors lead large NF1 services and manage children with NF1-associated OPG. The corresponding author is lead clinician in the National Commissioning Group Complex NF1 service at Guy's and St Thomas' NHS Foundation Trust, London.

References

1 Simmons I, Gogi D. Screening children with NF1 for optic pathway glioma—Yes. *Eye (Lond)* 2010; **24**(9): 1429–1431.
2 Pilling RF, Taylor RH. Screening children with NF1 for optic pathway glioma—No. *Eye (Lond)* 2010; **24**(9): 1432–1434.
3 Listernick R, Ferner RE, Liu GT, Gutmann DH. Optic pathway gliomas in neurofibromatosis-1: controversies and recommendations. *Ann Neurol* 2007; **61**(3): 189–198.
4 Grill J, Laithier V, Rodriguez D, Raquin MA, Pierre-Kahn A, Kalifa C. When do children with optic pathway tumours need treatment? An oncological perspective in 106 patients treated in a single centre. *Eur J Pediatr* 2000; **159**(9): 692–696.
5 Listernick R, Ferner RE, Piersall L, Sharif S, Gutmann DH, Charrow J. Late-onset optic pathway tumors in children with neurofibromatosis 1. *Neurology* 2004; **63**(10): 1944–1946.

DH Gutmann[1], R Listernick[2] and RE Ferner[3]

[1]Department of Neurology, Washington University School of Medicine, St Louis, MO, USA
[2]Division of General Academic Paediatrics, Feinberg School of Medicine, Northwestern University, Chicago, IL, USA
[3]Department of Neurology, Guy's Hospital and St Thomas' NHS Foundation Trust, London, UK
E-mail: rosalie.ferner@kcl.ac.uk

Eye (2011) **25,** 818; doi:10.1038/eye.2010.227; published online 28 January 2011

Sir,

Response to Gutmann *et al*

We are pleased that the publications have provoked open debate and thank Gutmann *et al* for their comments.[1]

We concur that the current recommendations of ophthalmology appointments for children with NF1 do not constitute screening, and should not be labelled as such. A recent audit of the NF1 OPG screening service in Manchester revealed that screening appointments were poorly attended and no OPGs were detected during annual ophthalmology screening over a 7-year, 128-episode period.[2]

The term 'age appropriate screening' remains problematic on several levels. At least 40% children with NF1 have developmental delay or learning disability and have difficulty performing the tests.[3] Learning disability is associated with cerebral visual impairment (CVI); CVI and amblyopia reduce vision. If there are no 'normal' data for visual development in children with NF1, how are we to define the 'reduced vision' that is 'highly predictive of OPG'?[1]

Perhaps what we should all be working towards is 'developmentally appropriate screening' and use the ongoing national project to collect the 'normal' data we need, so that we may be better informed when making the next round of recommendations.

Conflict of interest

The authors declare no conflict of interest.

References
1 Gutmann DH, Listernick R, Ferner RE. Screening for symptomatic optic pathway glioma in children with neurofibromatosis type 1 *Eye* 2011; **25**: 818.
2 Pilling RF, Lloyd IC, Huson SM. Utility of optic pathway glioma screening in young children with neurofibromatosis type I: questions generated by a clinical audit. *Eye* 2010; **24**(10): 1603–1605.
3 Krab LC, Aarsen FK, de Goede-Bolder A, Catsman-Berrevoets CE, Arts WF, Moll HA *et al*. Impact of neurofibromatosis type 1 on school performance. *J Child Neurol* 2008; **23**(9): 1002–1010.

RF Pilling[1] and RH Taylor[2]

[1]Bradford Royal Infirmary, West Yorkshire, UK
[2]York District Hospital, Yorkshire, UK
E-mail: rfpilling@btopenworld.com

Eye (2011) **25,** 818–819; doi:10.1038/eye.2010.230; published online 28 January 2011

Sir,

Brow suspension using 3-0 Prolene

We read with interest your publication of Garrott *et al*'s[1] recent case series, highlighting the long-lasting benefit of 2-0 Prolene brow suspension for congenital ptosis. Our experience of ~10 years of performing suture brow suspension in children and adults, for a number of different indications, echoes these findings. In addition, we found that repeat brow suspension, when necessary, was a more straightforward undertaking using this technique. This technique avoids the sequelae of autologous fascia lata harvesting[2] and the soft tissue complications associated with the use of mersilene mesh.[3]

A retrospective case note review highlighted 41 eyes having undergone brow suspension surgery using 3-0 Prolene over a 9-year period (2001–2009) at our institute. All cases with a levator function of <5 mm were included in the analysis, irrespective of cause of ptosis and age, to provide a realistic clinical picture. Mean age at operation was 23 years (range 1–74 years). Recorded palpebral aperture improved significantly following the surgery ($P<0.0005$; mean 4 mm pre-operation, 7 mm post-operation). The majority were free from complications, although seven (17%) experienced a recurrence of their ptosis requiring a further operation and five (12%) experienced discomfort or a lump at the site of the suture knot. The Kaplan–Meier plot shows the cumulative survival for the group (Figure 1).

Unfortunately, no randomized controlled trials have been undertaken to determine the optimal material for brow suspension surgery. We commend Garrott *et al* for publishing their long-term outcomes and agree that fascia lata brow suspension is not a prerequisite for successful, long-lasting brow suspension surgery in children. Indeed fascia lata use is not without its complications, at both the harvesting site[2] and through proposed contracture of the fascia lata itself leading to variability in the cosmetic results.[4] In addition, we suggest a Prolene sutured brow suspension can be used in a broader group of patients, to provide adults with ptosis and poor levator function, a cosmetically and functionally satisfactory result in the long-term. It is worth noting that we have performed this procedure under local anaesthesia in a small number of adults. It is an easily reversible and repeatable procedure.

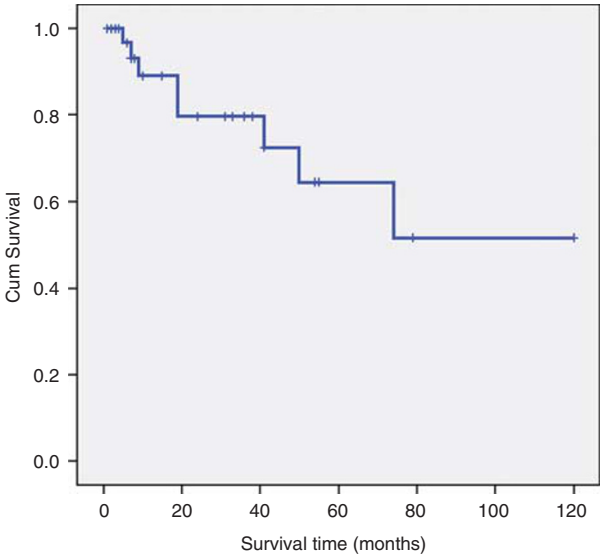

Figure 1 Kaplan–Meier plot demonstrating cumulative survival for brow suspensions using 3-0 Prolene.

Conflict of interest

The authors declare no conflict of interest.

References
1 Garrott H, Aristodemou P, Sinclair N, Lane C, Harrad R. Long-term efficacy of 2-0 Prolene brow suspensions for congenital ptosis. *Eye* 2010; **24**(1): 175–177.
2 Bleyen I, Hardy I, Codère F. Muscle prolapse after harvesting autogenous fascia lata used for frontalis suspension in children. *Ophthal Plast Reconstr Surg* 2009; **25**(5): 359–360.
3 Mehta P, Patel P, Olver JM. Functional results and complications of mersilene mesh use for frontalis suspension ptosis surgery. *Br J Ophthalmol* 2004; **88**(3): 361–364.
4 Yoon JS, Lee SY. Long-term functional and cosmetic outcomes after frontalis suspension using autogenous fascia lata for pediatric congenital ptosis. *Ophthalmology* 2009; **116**(7): 1405–1414.

AA Mokashi, RE Stead and LC Abercrombie

Department of Ophthalmology, Nottingham University NHS Trust, Queens Medical Centre, Nottingham, UK
E-mail: aamokashi@gmail.com

Eye (2011) **25,** 819; doi:10.1038/eye.2011.8; published online 11 February 2011