the considerations of the UK National Screening Committee about childhood screening for amblyopia.

We hope the very large number of ophthalmologists who support BOSU, including those who contributed specifically to our study, will be reassured about the quality and value of work undertaken through BOSU.

Far from employing 'suspect methodology', BOSU uses a well-established approach to provide a unique and powerful resource for the epidemiological study of uncommon ophthalmic disorders, which is envied outside the UK. The BOSU ensures that an evaluation of ascertainment is included in the study methodology and reported as part of the findings. In time, the studies undertaken through it can be expected to contribute a significant body of evidence on which clinical practice and policy will be based—as the example of the British Paediatric Surveillance Unit, now in its 17th year and on which BOSU is modelled, so clearly shows.[11] It would be a great pity if BOSU were prevented from fulfilling this potential role in ophthalmology in the UK.

## References

1 Thacker SB, Redmond S, Berkelman RL. A controlled trial of disease surveillance strategies. *Am J Prev Med* 1986; **2**: 345–350.
2 Vogt RL, LaRue D, Klaucke DN, Jillison DA. Comparison of an active and passive surveillance system of primary care providers for hepatitis, measles, rubella, and salmonellosis in Vermont. *Am J Public Health* 1983; **73**(7): 795–797.
3 Hook EB, Regal RR. The value of capture-recapture methods even for apparent exhaustive surveys. *Am J Epidemiol* 1992; **135**: 1060–1067.
4 International Working Group for Disease Monitoring and Forecasting. Capture–recapture and multiple-record systems estimation II: applications in human diseases. *Am J Epidemiol* 1995; **142**: 1059–1068.
5 International Working Group for Disease Monitoring and Forecasting. Capture–recapture and multiple-record systems estimation 1: history and theoretical development. *Am J Epidemiol* 1998; **142**: 1047–1058.
6 Rahi JS, Dezateux C, for the British Congenital Cataract Interest Group. Capture–recapture analysis of ascertainment by active surveillance in the British Congenital Cataract Study. *Invest Ophthalmol Vis Sci* 1999; **40**: 236–239.
7 Rahi JS, Logan S, Timms C, Russell-Eggitt I, Taylor DSI. Risk, causes, and outcomes of visual impairment after loss of vision in the non-amblyopic eye: a population-based study. *Lancet* 2002; **360**: 597–602.
8 Foot B, Stanford MR, Rahi JS, Thompson JR, on behalf of the British Ophthalmological Surveillance Unit. The British Ophthalmological Surveillance Unit: an evaluation of the first three years. *Eye* 2003; **17**: 9–16.
9 Evans J, Rooney C, Ashwood F, Dattani N, Wormald RPL. Blindness and partial sight in England and Wales: April 1990–March 1991. *Health Trends* 1996; **28**: 5–12.
10 Williams C, Harrard RA, Harvey I, Sparrow JM, the ALSPAC study. Screening for amblyopia in preschool children: results of a population-based randomised controlled trial. *Ophthalmic Epidemiol* 2001; **8**: 279–295.
11 Nicoll A, Lynn R, Rahi JS, Verity C, Haines L. Public health outputs from the british paediatric surveillance unit and similar clinician-based surveillance mechanisms. *J Roy Soc Med* 2000; **93**: 580–585.

J Rahi[1], M Stanford[2] and B Foot[2]

[1]Centre for Paediatric Epidemiology and Department of Ophthalmology, Institute of Child Health/Great Ormond Street Hospital, London, UK

[2]The British Ophthalmological Surveillance Unit, The Royal College of Ophthalmologists, London, UK

Correspondence: J Rahi, Centre for Paediatric Epidemiology and Department of Ophthalmology, Institute of Child Health/Great Ormond Street Hospital, 30 Guildord Street, London, UK
Tel: +44 020 79052250;
Fax: +44 020 7242 2723.
E-mail: j.rahi@ich.ucl.ac.uk

*Eye* (2005) **19,** 350–351. doi:10.1038/sj.eye.6701810
Published online 28 January 2005

---

Sir,

**Neurofibromatosis type 1 presenting with Horner's syndrome**

Johann Friedrich Horner[1] described the syndrome of ptosis, miosis, and anhydrosis as a result of interruption of sympathetic innervation to the eye in 1869. We describe a patient who presented with a preganglionic Horner's syndrome secondary to a malignant peripheral nerve sheath tumour who was subsequently diagnosed as having neurofibromatosis type 1 (NF1). This case highlights the importance of a thorough investigation of any patient presenting with a Horner's syndrome and, to the best of our knowledge, this is the first reported case of NF1 presenting with a Horner's syndrome.

### *Case report*

A 31-year-old woman presented with a 2-month history of a drooping left eyelid. She had no past ocular or medical history. There was a left-sided ptosis and pupil examination revealed an anisocoria that was greater in the dark. These findings were felt to be consistent with a left Horner's syndrome. Lisch nodules were noted

bilaterally (Figure 1). Optic discs were healthy bilaterally. Cutaneous examination revealed several small neurofibromas, some café-au-lait spots and axillary freckling. The remainder of the systemic examination was unremarkable and in particular there were no T1 physical signs to suggest a thoracic inlet (Pancoast's) syndrome.

A computerised tomography (CT) scan of the chest was performed (Figure 2), which showed a nonenhancing dumb-bell-shaped mass extending from the root of the neck anterior to the first rib to approximately 2 cm above the level of the aortic arch. The mass displaced the vessels in the root of the neck anteriorly and abutted the T2 vertebral body posteriorly. A further low attenuation left axillary mass was noted sitting inferolaterally to the left axillary vein and artery. The remainder of the mediastinum was normal and the lung parenchyma were clear.

The patient subsequently underwent exploration and resection of the tumours. The left axillary tumour was found to be a neural tumour in a small branch coming off the infraclavicular brachial plexus. The tumour in the root of the neck was discovered to be two separate tumours: one arising from the T1 nerve root and the other from the sympathetic chain. Histology showed that the axillary tumour was a plexiform neurofibroma and that the two tumours in the root of the neck were both malignant peripheral nerve sheath tumours (MPNST).

The patient was reviewed by the clinical geneticists who diagnosed NF1, on the basis that the three separate neural tumours combined with the cutaneous signs fulfilled the criteria established in 1988.[2] There was no previous family history of NF1. The patient made an excellent recovery from the operative procedure and is currently undergoing follow-up by the oncologists.

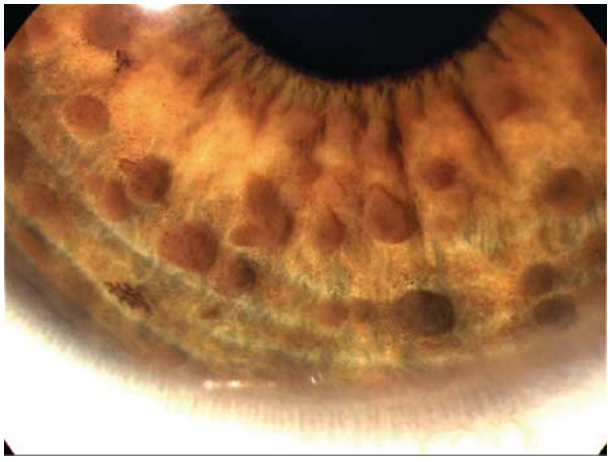

**Figure 1** Lisch nodules.

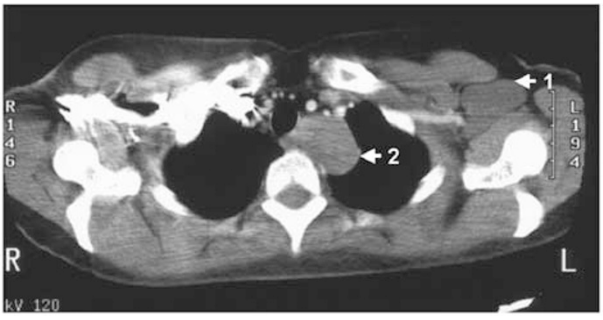


**Figure 2** Axial CT scan of the neck showing: (a) left axillary tumour (plexiform neurofibroma); (b) tumour in the root of the neck (MPNST).

## Comment

Patients with Horner's syndrome may be separated into three groups according to the site of the lesion, these being central, preganglionic, and postganglionic. [3] The preganglionic (second-order) neuron for sympathetic supply to the eye begins in the ciliospinal centre of Budge between the eighth cervical vertebra and the fourth thoracic vertebra (C8–T4) of the spinal cord. The axons exit the spinal cord via the anterior horn, pass through the pulmonary apex and enter the sympathetic chain in the neck, synapsing in the superior cervical ganglion. Although pharmacological testing of the patient's pupils was not performed, the pathology revealed as a result of surgery confirms the Horner's syndrome in this case to be preganglionic in origin. Malignancy has been reported to be the cause of about 25% of cases of preganglionic Horner's syndrome with the most common tumours being lung and breast cancer.[4]

MPNSTs are defined as soft-tissue tumours of presumed Schwann's cell lineage with anaplastic features in the form of high cellularity, cellular and nuclear pleomorphism, a high mitotic rate, and necrosis. A large number of terms have been applied to such lesions and include malignant schwannoma, malignant neurofibroma, malignant neurilemmoma, neurofibrosarcoma, and neurogenic sarcoma. The incidence of MPNST arising in NF1 is 4.6 and 0.001% in the general population.[5] Treatment consists of complete and wide surgical resection. Adjuvant irradiation seems to improve local control of the disease. The prognosis depends on tumour size, history of prior irradiation, surgical excision margin, and the histological presence of necrosis.[6] MPNSTs frequently recur locally and metastasize distantly, with the lung being the most common site of metastasis.

In most cases, MPNSTs present late with symptoms of enlarging mass and pain.[2,7] Our patient was fortunate that by presenting with a Horner's syndrome, further investigation resulted in the early diagnosis of MPNST. The subsequent prompt management of the MPNST will hopefully lead to a more favourable outcome in our patient.

## References

1 Horner F. Uber eine form von ptosis. *Klin Monatsbl Augenheilkd* 1869; **7**: 193–198.
2 Neurofibromatosis. Conference statement. National Institutes of Health Consensus Development Conference. *Arch Neurol* 1988; **45**: 575–578.
3 Maloney WF, Younge BR, Moyer NJ. Evaluation of the causes and accuracy of pharmacologic localization in Horner's syndrome. *Am J Ophthalmol* 1980; **90**(3): 394–402.
4 Thompson H, Maxner C, Corbett J. Horner's syndrome due to damage to the preganglionic neuron of the oculosympathetic pathway. In: Huber A (ed). *Sympathicus und Auge*. Ferdinand Enke: Stuttgart, 1990, pp. 99–104.
5 Bilgic B, Ates LE, Demiryont M, Ozger H, Dizdar Y. Malignant peripheral nerve sheath tumors associated with neurofibromatosis type 1. *Pathol Oncol Res* 2003; **9**(3): 201–205, (Epub October 15, 2003).
6 Kourea HP, Bilsky MH, Leung DH, Lewis JJ, Woodruff JM. Subdiaphragmatic and intrathoracic paraspinal malignant peripheral nerve sheath tumors: a clinicopathologic study of 25 patients and 26 tumors. *Cancer* 1998; **82**: 2191–2202.
7 Baehring JM, Betensky RA, Batchelor TT. Malignant peripheral nerve sheath tumor: The clinical spectrum and outcome of treatment. *Neurology* 2003; **61**(5): 696–698.

P Cackett, J Vallance and H Bennett

Princess Alexandra Eye Pavilion
Chalmers Street, Edinburgh EH3 9HA, UK

Correspondence: P Cackett
Tel: +44 0131 536 1674
Fax: +44 0131 536 1574
E-mail: pete@pdcackett.demon.co.uk

*Eye* (2004) **19,** 351–353. doi:10.1038/sj.eye.6701478
Published online 23 July 2004

Sir,
**Reply**

The authors of the letter made several points regarding the validity of our results. We agree that a correlation coefficient plot does not necessarily exclude systematic bias or disagreement between measurements obtained by the two methods being evaluated. This is the reason for quantifying agreement using the Bland–Altman graphical method.[1] In our paper we elected to use analogue measurements on the horizontal axis as this was regarded as the gold standard.[2] Re-plotting the graph using an average of analogue and digital on the horizontal axis did not make any difference to the limits of agreement.

We found that the limits of agreement for distance up to 5 mm were clinically acceptable, but we do accept that there appears to be a linear relationship between amount of disagreement and magnitude of distance measured. We are grateful to the authors for pointing this out, and would suspect that the most likely source of this bias might be the actual screen size (number of pixels) setting on the computer monitor. This would explain the similar gradient seen in group 1 and group 2 plots. We will conduct further studies to evaluate the influence of screen size setting as a confounding factor.

## References

1 Bland JM, Altman DG. Statistical methods for assessing agreement between two methods of clinical measurement. *Lancet* 1986; **1**(8476): 307–310.
2 Musadiq M, Patsoura E, Hughes S, Yang YC. Measurements of linear dimensions on fundus photographs: comparison between photographic film and digital systems. *Eye* 2003; **17**(5): 619–622.

M Musadiq and YC Yang

Wolverhampton Eye Infirmary
Compton Road Wolverhampton
WV3 9QR, UK

Correspondence: M Musadiq
Tel: +44 1902 645023
Fax: +44 1902 645018
E-mail: mmusadiq@hotmail.com

*Eye* (2005) **19,** 353. doi:10.1038/sj.eye.6701486
Published online 23 July 2004

Sir,
**Hemiretinal vein occlusion associated with pseudotumour orbit: an observational case report**

Pseudotumour orbit is a condition of idiopathic nonspecific orbital inflammation with associated retinal changes such as papillodema, papillitis, choroiditis, and