Congenital eye anomaly surveillance in England and Wales. How effective is the national system?

SP Shah^{1,2}, B Botting³, A Taylor¹, Y Abou-Rayyah², J Rahi^{4,5} and CE Gilbert¹

EDITORIAL

Eye (2011) 25, 1247–1249; doi:10.1038/eye.2011.207

Surveillance is defined as ‘the ongoing systematic collection, analysis, interpretation, and dissemination of data, reflecting the current status of a community or population’.¹

Congenital anomaly surveillance is important in improving the health of children,² and anomaly registers are well established in many high-income countries. The data collected can be used to detect geographical clustering of cases or temporal trends, both of which may indicate change in exposure to harmful environmental agents. The data also serve to inform service planning and medical/epidemiological research.

Routine reporting of surveillance data for congenital anomalies in England and Wales has been performed by the National Congenital Anomaly System (NCAS) run by the Office for National Statistics (ONS).³ NCAS notification³ systems were complex, but essentially, there were two tiers, (a) an original system of voluntary notifications (completed by any health professional) using the ‘SD56’ reporting form, and (b) contribution of data to the NCAS scheme by regional anomaly registers.⁴ Regional anomaly registers have been established at different times, with different purposes, and with different funding arrangements. They only started to exchange data with NCAS in 1998, and they are recognised as having more complete case ascertainment⁵ than the ‘SD56’ system.

Congenital eye anomalies are an important cause of visual impairment in children worldwide, and are responsible for approximately 15–20% of blindness and severe visual impairment in children.⁶ The majority of these anomalies have an unknown aetiology,

but both genetic and environmental factors have a role.^{7–9} The completeness in ascertainment of eye anomalies by NCAS has not been examined since the inclusion of regional register data. The purpose of this study was to compare the number of children identified in a national active surveillance study (ascertained through the British Ophthalmic Surveillance Unit (BOSU)¹⁰) with an equivalent NCAS data set. In the BOSU study, ophthalmologists reported all children aged 16 years, newly diagnosed with anophthalmos, microphthalmos, and/or coloboma (AMC). The NCAS data set included children with the International Statistical Classification of Diseases and Related Health Problems 10th Revision (ICD-10) codes applicable to eye anomalies (ie, Q10 to Q15¹¹). The two methods of case ascertainment were compared using a method described by Rahi and Botting.¹² The comparison was restricted to children born in England and Wales between 1 January 2007 and 31 December 2007, and four identifiers were used to match children. Each child in the BOSU data set was assigned one of four outcomes, three in which the child’s identifiers matched those in the NCAS data set, and one when no match was present (Table 1). Children in the NCAS data set, but who were not in the BOSU data set, were also identified.

The BOSU data set included 55 children, but only eight (14.5%) of them were listed in the NCAS data set as having an AMC code (ie, outcome 1; Table 1). The majority of children in the BOSU data set (81.8%) did not match any child in the NCAS data set. Eleven children were in the NCAS data set, but not in the BOSU data set.

Children with systemic anomalies were more likely to be on the NCAS register than those

¹International Centre for Eye Health, London School of Hygiene and Tropical Medicine, London, UK

²Moorfields Eye Hospital, London, UK

³Office For National Statistics, London, UK

⁴MRC Centre of Epidemiology for Child Health, Institute of Child Health, University College London, (UCL), London, UK

⁵Ulverschroft Vision Research Group, Institute of Child Health, UCL, London, UK

Correspondence: SP Shah, International Centre for Eye Health, London School of Hygiene and Tropical Medicine, London WC1E 7HT, UK.
Tel: +44 797 366 2745;
Fax: +44 207 958 8325;
E-mail: Shaheen.shah@lshtm.ac.uk

Meeting presentation: The Royal College Of Ophthalmologists Annual Congress 2009

Table 1 Results of matching the BOSU and NCAS data sets

Outcome		<i>n</i>	%
1	Identifiers matched and the child on the NCAS list had an AMC code	8	14.5
2	Identifiers matched, but the child on the NCAS list did not have an AMC code, but had another ICD-10 eye code (cataract for example)	0	0
3	Identifiers matched, but the child on the NCAS list had no ICD-10 eye code, but had a code of a systemic syndrome	2	3.6
4	No match: children not found on the NCAS list	45	81.8
5	No match: children only on NCAS list, and not in SEA-UK database	11	NA
Total		55	100

without (17.6 *vs* 9.5%). Children (8%) in the BOSU data set from voluntary notification areas (ie, 'SD56' system) were matched, compared with 20% from register regions. Two children identified through BOSU with anophthalmos, one bilaterally affected, were registered to NCAS as having systemic abnormalities, but no eye anomalies. All 11 children who were only in the NCAS register were recorded by regional registers.

This study shows that there is significant underreporting of eye anomalies to NCAS in England and Wales, with children living in an area served by a regional register being more likely to have been reported than those served by the SD56 system alone. Children with systemic anomalies were also more likely to be reported. This differential case ascertainment not only underestimates the number of children affected, but over emphasises the degree of severity.

Two other studies have explored completeness of reporting to NCAS of children with eye anomalies ascertained through other methods.^{12,13} Both studies were performed before the regional registers started contributing to NCAS, and so they only investigated the passive SD56 system. In the first study, only 22% of children with anophthalmos and microphthalmos identified using multiple sources between 1988 and 1994 were reported to NCAS.¹³ In the second study, only 10% of children newly diagnosed with cataract reported to BOSU between 1995 and 1996 were also reported to NCAS.¹² The low figures in this latter study and in ours may, in part, be due to more complete case ascertainment through BOSU, which uses active surveillance which is known to yield higher case ascertainment rates.¹⁴

Since the conclusion of this study, ONS has undertaken a consultation to consider the future of NCAS.¹⁵ Response to the consultation proposes that the SD56 system be terminated immediately, and discussions undertaken with the Department of Health for a properly funded, comprehensive national system based on existing regional registries that can provide good quality data on anomalies on both live births, still births, and terminations of pregnancy.¹⁶ We support and await the development of such a system; however, we are

concerned that, at present, only 50% of births in England are covered by a regional register.¹⁷

Eye anomalies can be very difficult to diagnose for the non-specialist, and an increased awareness of the importance and need to report cases by the paediatric eye health community is required. Greater engagement between coordinators of the regional registers and the paediatric eye care professionals would improve reporting, as would the designation of one member of staff in each paediatric eye unit to be responsible for day-to-day monitoring and reporting. Making the reporting mechanism more user-friendly, by allowing direct reporting by secure email, for example, may aid reporting by busy clinicians.

For the future, the increasing use of computerised medical records allowing linkage of databases is likely to change the face of public health surveillance, but for the present, national anomaly reporting for eye anomalies remains insensitive to detect changes in frequency and therefore risk factors.

Conflict of interest

JR is a member of the Ulverscroft Vision Research Group, and is based at Great Ormond Street Hospital/UCL Institute of Child Health, which receives a proportion of its funding from the Department of Health's National Institute of Health Research (NIHR) Biomedical Research Centres funding scheme. JR is based in the Medical Research Council's Centre of Epidemiology for Child Health and holds a joint appointment with Moorfields Eye Hospital/Institute of Ophthalmology, UCL, which also received a proportion of its funding from the NIHR Biomedical Research Centres funding scheme. All other authors declare no conflict of interest.

Acknowledgements

This study was supported by an unrestricted grant from Moorfields Special Trustees, and from Newlife Foundation. The funders had no role in the study design, data collection, data analysis, manuscript preparation, and/or publication decisions of this paper.

References

- 1 Thacker SB, Berkelman RL. Public health surveillance in the United States. *Epidemiol Rev* 1988; **10**: 164–190.
- 2 Wilcox AJ. The health of our children. *Am J Epidemiol* 1999; **150**: 665–666.
- 3 Office of National Statistics. *Congenital Anomaly Statistics*. <http://www.ons.gov.uk/ons/search/index.html?newquery=Congenital+Anomaly+Statistics>. Newport; 2008.
- 4 OPCS. The Office for Population Censuses and Surveys monitoring scheme for congenital malformations occasional paper 43. A review by a working group of the registrar general's medical advisory committee. 1995.
- 5 Misra T, Dattani N, Majeed A. Evaluation of the National Congenital Anomaly System in England and Wales. *Arch Dis Child Fetal Neonatal Ed* 2005; **90**: F368–F373.
- 6 Gilbert C, Foster A. Childhood blindness in the context of VISION 2020—the right to sight. *Bull World Health Organ* 2001; **79**: 227–232.
- 7 Morrison D, FitzPatrick D, Hanson I, Williamson K, van Heyningen V, Fleck B *et al*. National study of microphthalmia, anophthalmia, and coloboma (MAC) in Scotland: investigation of genetic aetiology. *J Med Genet* 2002; **39**: 16–22.
- 8 Verma A, FitzPatrick D. Anophthalmia and microphthalmia. *Orphanet J Rare Dis* 2007; **2**: 47.
- 9 Gregory-Evans CY, Williams MJ, Halford S, Gregory-Evans K. Ocular coloboma: a reassessment in the age of molecular neuroscience. *J Med Genet* 2004; **41**: 881–891.
- 10 Shah SP, Taylor AE, Sowden JC, Ragge NK, Russell-Eggitt I, Rahi JS *et al*. Anophthalmos, microphthalmos, and typical coloboma in the United Kingdom: a prospective study of incidence and risk. *Invest Ophthalmol Vis Sci* 2011; **52**: 558–564.
- 11 World Health Organization. *ICD-10 International Statistical Classification of Diseases and Related Health Problems* 10th Revision. 1994.
- 12 Rahi JS, Botting B. Ascertainment of children with congenital cataract through the National Congenital Anomaly System in England and Wales. *Br J Ophthalmol* 2001; **85**: 1049–1051.
- 13 Busby A, Dolk H, Collin R, Jones RB, Winter R. Compiling a national register of babies born with anophthalmia/microphthalmia in England 1988–94. *Arch Dis Child Fetal Neonatal Ed* 1998; **79**: F168–F173.
- 14 Thacker SB, Redmond S, Rothenberg RB, Spitz SB, Choi K, White MC. A controlled trial of disease surveillance strategies. *Am J Prev Med* 1986; **2**: 345–350.
- 15 ONS. <http://www.ons.gov.uk/ons/about-ons/consultations/closed-consultations/2010/consultation-on-the-future-of-the-national-congenital-anomaly-system/index.html>. London, 2010.
- 16 Office of National Statistics. The Future of the National Congenital Anomaly System - Response to the consultation. <http://www.ons.gov.uk/ons/about-ons/consultations/closed-consultations/2010/consultation-on-the-future-of-the-national-congenital-anomaly-system/index.html>. Accessed April 2011.
- 17 Boyd PA, Armstrong B, Dolk H, Botting B, Pattenden S, Abramsky L *et al*. Congenital anomaly surveillance in England—ascertainment deficiencies in the national system. *BMJ* 2005; **330**: 27.