must be established, both in terms of increased risk of bilateral visual impairment and in terms of quality of life. Second, the total cost of screening and treatment, and the actual benefit of a screening programme to the population,^{12,13} must also be assessed. As of today, we do not have all of the information required, but van Leewen *et al.*'s work has provided us with very valuable data, moving us closer to determining whether preschool vision screening can be justified.

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Congenital rubella

Congenital rubella syndrome: the end is in sight

Scott R Lambert

Rubella vaccination programmes offer hope in eradicating congenital rubella syndrome in developing countries

Rubella is usually a mild febrile illness associated with a rash, lymphadenopathy, conjunctivitis, malaise and arthralgia. In 1941, an Australian ophthalmologist, Norman Gregg, first reported the association between congenital cataracts and a rubella infection during pregnancy in an article entitled "Congenital cataract following German measles in the mother".¹ This condition is now referred to as congenital rubella syndrome (CRS). It occurs as a result of a maternal rubella infection during the first trimester of pregnancy when organogenesis is occurring and is most commonly associated with hearing impairment (60%), congenital heart disease (45%), microcephaly (27%) congenital cataracts (25%) and mental retardation (13%). During the early 1960s, there was a worldwide pandemic of CRS. In the United States alone, 20 000 children were born with CRS between 1962 and 1965. In 1969, a live attenuated rubella vaccine was licensed in the US, and widespread vaccination of children was initiated. As a result, CRS has almost been eradicated from the US. Between 2001 and 2004, there were only 4 reported

cases of CRS in the US, of which 3 were born to mothers who had immigrated to the US.²

In 2003, the Pan American Health Organization set a goal of eradicating rubella from the Western Hemisphere by 2010. Forty-three of the 44 member countries of the Pan American Health Organization have now instituted rubella vaccination programmes.³ With childhood vaccination programmes, the first dose is usually given at age 12–15 months and the second dose either at the time of school entry or in the early teen years. While all industrialised countries now have rubella-vaccination programmes, less than one-half of developing countries have rubella-vaccination programmes. Neither of the two most populous countries in the world, China and India, with a combined population of 2.5 billion people, has a rubella vaccination programme. It is estimated that 100 000 children are born each year in the world with the CRS.⁴ In order for a childhood vaccination programme to be successful, more than 80% of children need to be vaccinated in order to achieve herd immunity. It has been recommended by the World Health

Organization (WHO) that women of childbearing years also be vaccinated when a childhood vaccination programme is initiated in a country. Inadequate childhood vaccination programmes are believed to actually increase the risk of contracting rubella among women of childbearing years. In countries with childhood vaccination programmes, the seronegativity rate for rubella among women of childbearing years can be as low as 1%, whereas in countries without vaccination programmes, the seronegativity rate can be as high as 50%.^{5–10}

In India, CRS is the second leading cause of non-traumatic childhood cataracts exceeded only by hereditary cataracts.¹¹ The visual outcome of children with cataracts secondary to CRS is generally quite poor. In a recent report from India, 50% of children with bilateral cataracts secondary to CRS remained legally blind following cataract surgery.¹² The poor visual outcomes were ascribed to amblyopia, glaucoma, cornea opacities, optic atrophy and pupillary membranes. The poor visual outcomes were particularly tragic, since more than 50% of these children had a hearing impairment.

In its surveillance of CRS, the WHO distinguishes between: suspected cases of CRS, clinically confirmed cases of CRS and laboratory confirmed cases of CRS.¹³ The WHO defines a suspected case of CRS as an infant less than 1 year of age with heart disease and/or suspicion of deafness and/or one or more of the following ocular findings: leucocoria, decreased vision, nystagmus, strabismus, microphthalmos or buphthalmos. WHO defines a clinically confirmed case of CRS as an infant with two or more of the following findings confirmed by a physician: cataract, congenital glaucoma, pigmentary

retinopathy, congenital heart disease, hearing impairment or an infant with one of the findings listed above and one of the following findings: purpura, splenomegaly, microcephaly, mental retardation, meningoencephalitis, radiolucent bone disease or neonatal jaundice. A laboratory-confirmed case is an infant with clinically confirmed CRS who has rubella-specific IgM antibodies in their blood.

The Aravind Eye Hospital in Madurai, India has developed community-based screening for ocular signs as a means of identifying suspected cases of CRS. In this issue of the BJO, Vijayalakshmi and co-workers¹⁴ report screening of over 50 000 children for CRS (see page 1467). Nearly 2% of the children screened had ocular signs suggestive of CRS, and 27% of the children with suspected CRS were later found to have clinically confirmed CRS. They report that a cataract had the highest sensitivity (73%) for detecting CRS, and pigmentary retinopathy and iris hypoplasia had the higher specificity for CRS. Tragically, only 6 of the nearly 992 mothers of children with suspected CRS had been vaccinated against rubella. Clearly, CRS is still an enormous public health problem in India.

The WHO has proposed three stages to eliminate CRS from countries lacking rubella vaccination programmes.¹⁵ The first stage is to investigate rubella outbreaks, to assess the number of CRS cases per year for at least 2 years, and to assess the proportion of women at risk for

rubella during pregnancy. The second stage is to begin a national rubella immunisation programme, to report data on suspected rubella cases on a monthly basis and to investigate each suspected case of CRS. The third stage is to investigate and report every case of a febrile rash illness within 48 h. Hopefully, as developing countries find the necessary resources to initiate effective rubella vaccination programmes in the near future, the scourge of blindness and deafness arising from CRS can be eradicated from this planet.¹⁵

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Cionni capsular tension ring implantation in Marfan's syndrome

Cionni capsular tension ring implantation in Marfan's syndrome

Thomas S Dietlein

Progress is being made in dealing with Marfan's syndrome, but careful surgical planning decisions are still necessary

Safe positioning of an intraocular lens (IOL) after lentectomy in hereditary ectopia lentis has been achieved by various techniques including iris fixation by clipping or suturing, anterior chamber IOL with chamber-angle support and transscleral suturing of a posterior chamber IOL. None of these approaches requires preservation of the capsular bag, and they are regularly combined

with an anterior vitrectomy. However, the loss of separation between the anterior and posterior compartment of the eye may make a key contribution to subsequent complications such as retinal detachment, macular oedema or even glaucoma. From an anatomical point of view, the surgical approach proposed by Cionni *et al*^{1,2} and now studied by Bahar *et al*³ (see page 1477), in a series of adult

patients with hereditary lens subluxation owing to Marfan's syndrome, would seem to represent progress compared with traditional lentectomy with anterior vitrectomy and implantation of an anterior chamber lens. The implantation of the IOL in the transsclerally fixated capsular bag does not compromise the endothelial cell count over the years and should also cause less chronic anterior chamber irritation than is induced by the different anterior chamber IOLs. The preservation of an anterior and posterior compartment should also reduce the probability of retinal problems and make any subsequent glaucoma or vitreoretinal interventions easier, although naturally no sufficiently valid long-term data are available as yet.

However, some limitations of this very demanding surgical approach should be borne in mind by the interested cataract surgeon. In Marfan's syndrome, the anterior continuous curvilinear capsulorhexis may be complicated by the