phg hna toolkit | Congenital Disorders

Background information on Congenital Rubella Syndrome and the impact of interventions

This document gives a brief overview about the condition, its epidemiology and specific interventions that may reduce disease burden. The WHO has extensive information relating to rubella surveillance and immunisation which can be accessed via their website.

What is rubella?

Rubella is a viral disease directly transmitted through airborne droplets from person to person when coughing or sneezing. In susceptible populations, it commonly occurs during childhood, but can present at any age as a self-limited rash, low-grade fever, malaise and mild conjunctivitis. It has an incubation period of 14-21 days with the majority of infected individuals developing a rash 14-17 days after exposure. Individuals remain infectious from 7 days before symptoms appear to 4 days after the onset of the rash. Although rubella is normally a mild childhood disease, infection in pregnant women can have serious consequences for the fetus.

Rubella in pregnancy and Congenital Rubella Syndrome

Rubella infection during pregnancy can lead to fetal loss or Congenital Rubella Syndrome (CRS) resulting from mother-to-child vertical transmission of the infection. CRS occurs in up to 90% of infants born to women who acquire rubella during the first trimester and in the first 10 weeks of pregnancy in particular; the risk decreases to 10 to 20% between 11 and 16 weeks, and is very low beyond the second trimester.

CRS is an important cause of many severe birth defects: it is most commonly associated with intra-uterine growth retardation, microcephaly, hearing impairment, congenital heart diseases, congenital cataracts, other eye abnormalities and learning disabilities. Some infants may appear normal at birth, especially when infection occurred beyond the first trimester of pregnancy, but can develop sight problems or perceptive deafness.

What are the main risk factors?

Unvaccinated pregnant women who have not had rubella are at risk, especially when there are high levels of virus circulating in the population; this is more likely in places without universal vaccination for rubella or if vaccination coverage is low.

Global epidemiology

There is significant regional variation in the incidence of rubella and CRS, which is related to the impact of screening and immunisation programmes. In areas where the rubella vaccine has not been introduced into national immunisation programmes, rubella has a history of occurring at endemic and epidemic levels every 5-9 years. Figure 1 illustrates coverage of rubella vaccination. By the end of 2011, 131 countries routinely administered the rubella vaccine. Universal rubella vaccination has led to a remarkable reduction in cases, e.g. a 95% reduction in rubella and confirmed cases of congenital rubella in the Americas. Globally in 2010 there were 112,265 reported cases of rubella, and 208 cases of CRS[1]. However, these figures grossly underestimate the true population incidence. In developing countries the incidence of CRS may reach 0.5-3.3 per 1,000 live births during epidemics[2].

Surveillance data (Table 1) from Africa, the Americas, Asia, Eastern Europe, and the Eastern Mediterranean have recorded incidence rates of CRS of between 0.4 and 4.3 per 1,000 live births in epidemic years. The birth prevalence in non-epidemic years tends to be lower, but is usually severely under-estimated.

Surveillance of CRS is not well documented, but more than 100,000 cases are estimated to occur annually worldwide[3]. Surveillance reports show that the greatest number of cases are in Europe and the Americas; however, this reflects the better reporting and surveillance in these regions. The WHO has developed guidelines on developing improved CRS and rubella surveillance and monitoring systems as well as standard case definitions in order to improve reporting.

Mortality

The global mortality from congenital rubella has dropped substantially since the introduction of rubella vaccination, which was licensed in 1969. Nevertheless, the life expectancy of those born with congenital rubella syndrome is reduced, depending on disease severity, and access to and quality of health services.

Disability and quality of life

CRS is disabling e.g. through blindness (25% of cases), deafness (60%), congenital heart disease (45%) and learning disability (13%)[4]. The severity of CRS is variable, as is the amount of care and management that is required. Identification of cases in the prenatal and neonatal periods can reduce morbidity associated with CRS through effective support and counselling to the infected mother and affected child.

Reducing prevalence, morbidity and mortality

Global strategies to eliminate CRS are based on population immunisation and screening programmes. Population immunisation strategies may vary and will have to be decided by individual countries based on an assessment of what is feasible. Screening before and during pregnancy to assess women's immunity to rubella also helps informs action, including vaccination (before pregnancy), and gives information on risks during pregnancy or reassurance for those who are immune.

[1]WHO vaccine-preventable diseases: monitoring system 2012 global summary
[2]Robertson SE, Featherstone DA, Gacic-Dobo M, Hersh BS *Rubella and congenital rubella syndrome: global update*. Rev Panam Salud Publica. 2003 Nov;14(5):306-15
[3]ibid
[4]WHO The immunological basis for immunization series: module 11: rubella. 2008 ISBN 978 924 159684 8

Appropriate identification and treatment following birth and throughout life are also important elements of care for individuals affected by CRS. Figure 2 illustrates some of the opportunities for interventions.

Interventions before pregnancy

Universal immunisation programmes as part of national health programmes decrease the number of susceptible individuals and the rate of transmission of infection. Rubella vaccination gives immunity in 95% of those vaccinated after one dose and 99% after 2 doses[5]. There are different vaccination strategies to reduce the incidence of congenital rubella syndrome (CRS) or eliminate rubella, and the WHO has produced a number of documents in relation to immunisation and surveillance programmes for rubella.

Ideally all girls should have been vaccinated against rubella by the time they achieve reproductive age. Programmes should aim to achieve coverage of at least 95% in children (girls and boys); lower coverage could reduce transmission in childhood but leave a greater number of susceptible women at reproductive age, thus increasing the risk of rubella in pregnancy and CRS.

In places where rubella vaccination is not routinely given (including where single measles vaccine is provided), introducing catch-up programmes (targeting broader age groups) using measles, mumps and rubella (MMR) or measles-rubella (MR) vaccines may be recommended. However, as with above, this requires ensuring that immunisation coverage is high (>80%). Consideration of existing immunisation programmes for other diseases is an important aspect of developing a rubella immunisation strategy.

Interventions during pregnancy

A pregnant woman's risk of contracting rubella can be assessed by screening to check her immune status. If susceptible, they may be advised to avoid contact with people with rubella and, once the baby is born, they can be immunised against rubella to protect future pregnancies.

Clinical diagnosis of rubella, including in pregnancy, is unreliable as a large number of cases can be sub-clinical and symptoms are often non-specific. If a pregnant women develops symptoms and signs of a rubella-like illness or has been exposed to rubella, her susceptibility and gestational age should be determined. If she is shown to be immune to rubella or in advanced pregnancy, no further testing may be required. Otherwise laboratory confirmation of rubella may be required. Ultrasound may detect abnormalities in an infected fetus, but sensitivity may be low.

Susceptible pregnant women exposed to rubella should be counselled about the risk of vertical transmission, which varies inversely with gestational age, and offered management options such as the possibility of pregnancy termination (where this option is legal and acceptable), especially if primary infection has occurred before 12-16 weeks gestation. If termination of pregnancy is illegal or unacceptable, counselling and assistance are required to prepare the woman or couple for the birth of a potentially disabled child. Human normal immunoglobulin is probably ineffective in avoiding congenital rubella, and should only be considered where termination of pregnancy is not acceptable.

[5] www.dh.gov.uk/en/PublicHealth/immunisation/Greenbook/index.htm

Interventions after birth

Supportive care and follow-up are key elements for the management of CRS. Treatment depends on clinical manifestations, which can develop and progress over time. Investigations to determine the severity of CRS may include neurologic, cardiac, ophthalmologic and audiologic examinations, and care may need to be multi-disciplinary.

Cost-effectiveness of interventions

There has been great progress towards the elimination of rubella and CRS in many countries. The cost of eliminating these conditions has been estimated to be 7% of what it would cost some countries' health systems to provide care and rehabilitation services for babies born with CRS (the reported cost of treating CRS ranges from $1,994 – $43,920). Studies examining the cost-effectiveness of rubella vaccination are limited and most have been conducted in high income countries, however, where they indicate that vaccination programmes are cost-effective.

Cost-effectiveness usually varies internationally. For cost-effectiveness cut-off points for different regions of the world, go to: http://www.who.int/choice/costs/CER_levels/en/index.html, and for costs for specific items by region and county, go to: http://www.who.int/choice/costs/en/.

What are the main ethical, legal and social issues (ELSI) to consider?

Issues such as equity, availability and accessibility of services, alongside legal issues, such as those related to pregnancy termination, may be relevant.

Preconception care

It is important that all those at risk should be able to benefit from vaccinations, screening for rubella susceptibility and other preventive interventions. Availability and accessibility of services vary across countries and regions, and programmes and their delivery are vulnerable to lack of funding, poor health service infrastructure, and local social and cultural backgrounds.

During pregnancy

In many countries termination of a pregnancy is illegal or severely restricted to cases where it is necessary to protect the woman's life. Termination may also be dependent on consent from a woman's parents or partner. Illegal abortions are common in places with poorer health care and infrastructure. This may lead to unsafe abortion practices and increased risk to the woman.

Living with a disability

Congenital rubella is a major worldwide cause of preventable hearing impairment, blindness and cognitive disability. Those who are born with congenital rubella often have a low life expectancy due to poor health care services, weak infrastructure, and often insufficient priority given to people with disabilities. Lack of access to and knowledge of available interventions also adds to low life expectancy.

KEY REFERENCES

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RELATED TOPICS

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Figure 1: Global coverage of rubella immunisation

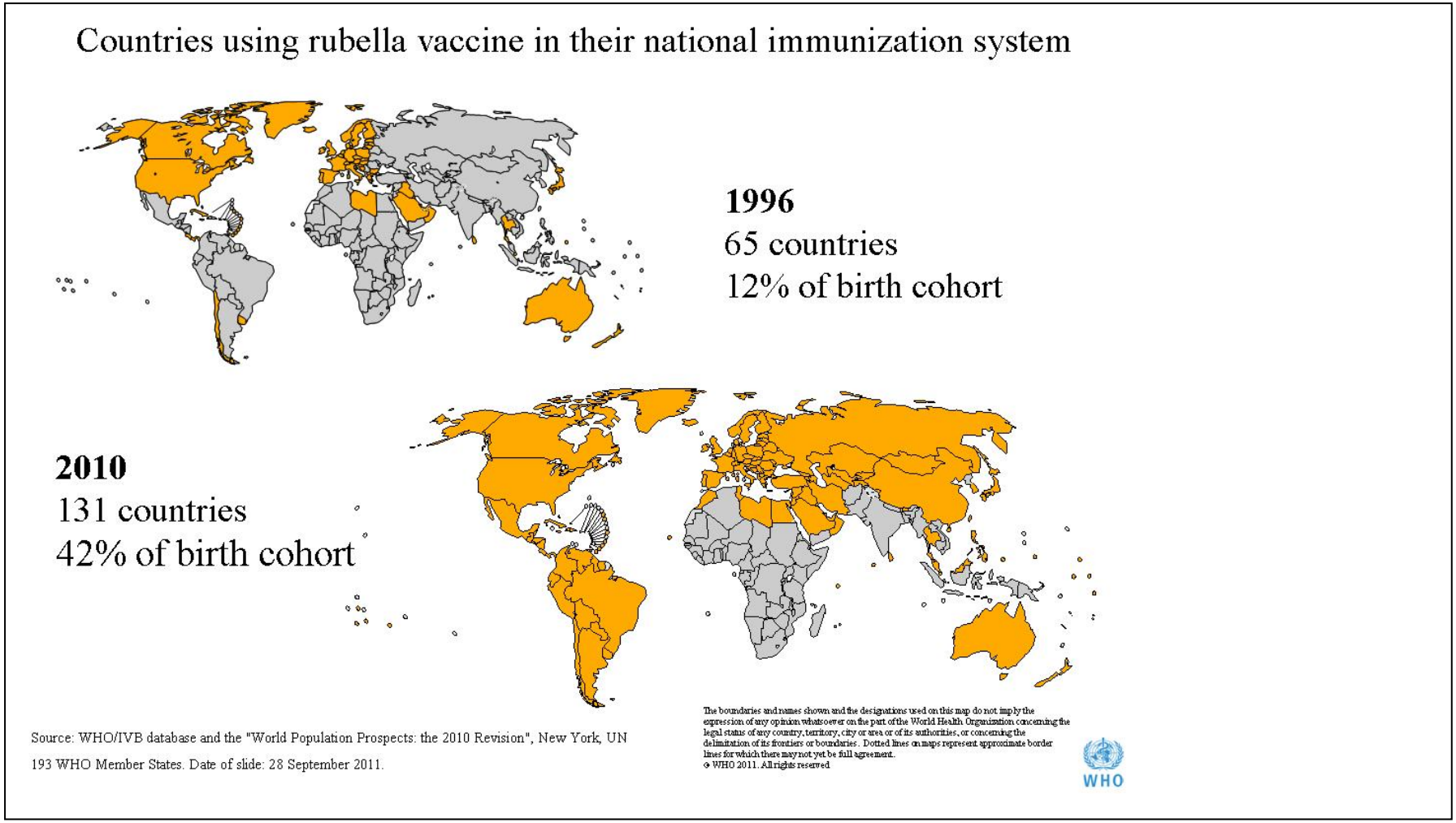

Table 1: Examples of reported incidence rates for CRS (from www.whoint/immunization/rub_global_review_2003.pdf)

Country	Place	CRS Incidence & Time Period
Brazil	City of Rio Branco	0.6/ 1000 live births in 2000-2001 outbreak, with 4.3/1000 live births in peak month
Ghana	Ashanti Region	0.8/ 1000 live births in 1996-1997
Israel	National	1.7/ 1000 live births in 1972
Jamaica	Nationwide institutions of handicapped children	0.4/ 1000 live births in 1972-1981
Jamaica	National	0.4/ 1000 live births in 1995 outbreak
Malaysia	University Hospital, Kuala Lumpur	0.5/ 1000 live births in 1993-1998
Oman	National	0.5/ 1000 live births in 1988 outbreak
Oman	National	0.7/ 1000 live births in 1993 outbreak
Panama	Panama City	2.2/ 1000 live births in 1985-1987
Russian Federation	Perm Region	3.5/ 1000 live births in 1994-1995

Figure 2: Needs assessment flowchart for congenital rubella