# Congenital Rubella Syndrome in Australia

Information Sheet

Congenital rubella syndrome (CRS) occurs in up to ninety per cent of babies born to women who are infected with rubella during the first ten weeks of their pregnancy. It can cause a miscarriage or anomalies of the heart and brain and can cause deafness and blindness.

Due to vaccination, incidence in Australia is now very low. Those with CRS, born during the rubella epidemic, are now in their forties or older, many of whom require frequent review by a range of medical specialists, highly trained carers and specialist supports for communication, orientation and mobility. During this era, CRS was one of the major causes of congenital deafblindness.

With well-matched services, people with CRS lead active lives and are involved in work and community life.



ID: A photo of Deb sitting on a stool in front of a green screen in a film studio, holding her script.

# Deb

My name is Deborah Kazich. I was born in 1971 with rubella syndrome. My rubella came from my mum having German measles when she was pregnant with me.

I have had hearing loss since I was born and I cannot hear without my hearing aids. My left ear works better than my right ear.

I had my sight until 2008 and I have worn glasses with thick lenses my whole life. In 2008 I had lots of surgeries on my eyes. In 2008 I also lost my sight in my right eye, so my left side is my best side for seeing and hearing. I use a white cane to help me navigate my community.

I live in my own home that I pay my mortgage on. I have worked with Good Sammy’s for 15 years now.

I enjoy spending time with people from the deafblind community. I love going for walks and since 2020 I have been going to Toastmasters each week to improve my public speaking skills so I can keep sharing awareness of my disability.

My favourite thing ever is craft - I dedicate a whole day on it. I really enjoy ceramics and mosaics as it helps me to feel relaxed and I like it because I like to challenge myself. I have even won awards for my mosaics and ceramics at the Canning Show.

I have recently started ballroom dancing again. I did ballroom dancing from 1988 to 2006, I stopped because there was no dancing school for the blind. I have recently found Ballroom Fit, where I do lessons each week and have monthly exams. I have won medals in three state competitions this year! I really love being back at dancing and it makes me feel proud.

Despite Rubella taking so much from me, I’ve been able to live a really exciting and fulfilling life.

This story was told by Deb, with help from support worker Cassie, to write it down.

# Incidence in Australia

In the 1940s, Australian ophthalmologist Sir Norman McAlister Gregg was the first to describe the connection between rubella infection in mothers, and cataracts and other birth anomalies in babies. This prompted the development of the rubella vaccine in the 1960s (Gidding, Dey & Macartney, 2018).

The estimate of the average number of births of infants with congenital rubella in Australia was 200 each year, between 1968 and 1976. This had dropped to less than 20 cases annually, by 1977 (Menser, Hudson, Murphy & Laurence, 1985).

An Australian doctor recognised the link between rubella infection in pregnant mothers and blindness in their babies. This discovery prompted the development of the rubella vaccine.

In 1970 the Rubella vaccine was approved and school girl vaccinations began in 1971. In 1989, via the Australian National Immunisation Program, rubella vaccination was available for all children (Gidding, Burgess & Kempe, 2001).

Today vaccination for rubella occurs at 12 months and 18 months of age (Australian Government Department of Health, 2020), giving a high level of protection against the disease (Australian Institute of Health and Welfare, 2018).

While vaccination has almost eradicated rubella in Australia there is still a risk of infection in the community. Groups at risk include

* those born overseas who may not have received rubella vaccines
* Aboriginal and Torres Strait Islanders from rural and remote communities
* and possibly, due to declining immunity over time, those aged 35 and over

*In Australia, there is still a small risk that a child will be born with Congenital Rubella Syndrome. Women considering pregnancy should be tested for rubella immunity.*

Between 1993 and 2013 there were 34 cases of children born with Congenital Rubella Syndrome, reported to the Australian Paediatric Surveillance Unit. Clinical characteristics of these children included deafness (13/34), congenital heart disease (12/34), cataracts (11/34), developmental delay (7/34) and retinopathy (4/34). Other features included microcephaly, celery stalking, intracranial calcification, thrombocytopenia, cerebral palsy, conjugated hyperbilirubinemia, pulmonary hypertension and hypoxic-ischaemic encephalopathy. Not all children had all features (Khandakera, Zurynskic & Jonesa, 2014).

# Situation in Australia

As a result of the rubella epidemic of the 1960’s and 70’s, the greatest number of Australians with Congenital Rubella Syndrome are currently aged between 40 and 60 years old.

Australians with Congenital Rubella Syndrome are mostly aged between forty and sixty years old

Information on this group is not widely documented (Simons, Reef, Cooper, Zimmerman & Thompson, 2014).

# Deafblindness in CRS

If a person has a combination of vision and hearing difficulties that affect communication, socialisation, mobility and daily living, they meet the criteria for deafblindness.

No studies could be found that reported the incidence of deafblindness in Congenital Rubella Syndrome. Studies in this area focus on the incidence of various eye conditions and hearing loss in people with deafblindness already identified.

Given the high incidence of eye conditions and deafness in CRS, deafblindness is a possible outcome for many

Characteristics of CRS affecting vision include microphthalmia, retinopathy, congenital cataracts, chorioretinitis, pigmentary and congenital glaucoma. Hearing can be affected by sensorineural deafness (Bourcoiran & Castillo, 2018).

In addition to the above, the following conditions affecting vision have also been reported in people with deafblindness with CRS; corneal clouding, kerataconus, corneal hydrops, cataracts, spontaneous lens absorption, glaucoma, diabetic retinopathy, retinal detachment, uveitis, strabismus, nystagmus, tear duct abnormalities and removal of one or both eyes (usually due to disease or pain).

(American Academy of Ophthalmology, n.d.; O'Donnell, 1991; Arnold, McIntosh, Martin & Menser, 1994; Canadian Deafblind Association (National), 2014; Gupta, Ali & Naik, 2017).

As many as half of those with CRS could have eye conditions affecting vision (Arnold, McIntosh, Martin & Menser, 1994).

Hearing loss is one of the most common characteristics of Congenital Rubella Syndrome.

A West Australian study conducted in 1985 reviewed 91 cases of children with CRS, born between 1968 and 1976. Results indicated nearly 70% had hearing loss ranging from mild to profound. It was thought that the prevalence of deafness may have been underestimated in this data due to some children’s deafness not being identified until 2 to 3 years of age to as late as school age (Stanley, Burgar, Fong & Milroy, 1985).

Given the high prevalence oculopathy and even higher prevalence of hearing loss in congenital rubella syndrome, the possibility of deafblindness should always be considered. This evaluation should be revisited periodically through the lifespan, as deterioration of both senses is reported in CRS literature.

This population has unique problems with communication, mobility and other daily living skills that make independent living more difficult to achieve. Additionally, many residential services have little or no experience providing the supports required by these adults (Armstrong, 2015).

# ****Health Implications****

The risk of CRS after maternal infection is greatest during the first sixteen weeks of pregnancy. There is little risk beyond twenty weeks of pregnancy, with intrauterine growth restriction seeming to be the only consequence of infections occurring in the last twelve weeks of pregnancy.

Delayed manifestations of CRS are symptoms or diseases with onset later in life, but are still directly or indirectly caused by the rubella infection and the resultant damage to the embryo. Delayed manifestations reported include (Dammeyer, 2010).

* Diabetes: overt, latent diabetes or abnormal insulin response, due to viral attack of pancreas cells
* Thyroid diseases, hypothyroidism, hyperthyroidism, and thyroiditis, due to autoimmune mechanisms
* Early menopause and osteoporosis, due to endocrine abnormalities
* Cardiovascular effects, hypertension, obstructive arterial lesions, aortic valve sclerosis
* Ocular damage as late-onset disease or as a progression of the congenital damage including glaucoma and spontaneous lens absorption
* Auditory damage, emergent late-onset deafness after the first year of life
* Growth hormone deficiency
* Psychosocial challenges, including anxiety attacks, anger, crying and self-injury;
* Progressive rubella panencephalitis (PRP): a name for a slowly progressive and fatal disease of the central nervous system that is due to chronic infection of the brain from rubella virus. Symptoms of PRP commonly develop in the second decade of life and may include learning problems, ataxia, nystagmus and cognitive decline with progression to seizures and cerebral palsy (Sever, South & Shaver, 1985).
* Gastrointestinal symptoms reported include cyclic vomiting, gagging, swallowing difficulties, oesophageal stricture, reflux and constipation. Modified and special diets are in place for some.
* Musculoskeletal conditions reported include scoliosis, kyphosis, osteoporosis and arthritis (Canadian Deafblind Association (National), 2014).
* Hypoplastic tooth enamel has been reported in people with CRS, making their teeth more susceptible to tooth decay. Difficulty achieving adequate oral hygiene and teeth cleaning is also reported (Bhatia, Goyal, Dubey, Kapur & Ritwik, 2012). Specialist dentistry with sedation may be required (Ahuja, Shigli, Thakur & Jain, 2015).

# Autism and CRS

Reported rates of autism in children with CRS from several studies conducted in the late 60’s and early 70’s during the rubella epidemic, range from 7.4 % to 12.5%. This was over 200 times higher than population-based rates at the time. Multiple theories regarding the link have been proposed, including viral attack on the nervous system, and vitamin A toxicity (Mawson & Croft, 2019).

Others recognise that people with congenital deafblindness may struggle acquiring skills in social interaction and communication, developing theory of mind and may display repetitive behaviours and seemingly unusual responses to their environment. They may also become anxious or upset by changes in their environment and routine. All these observations could be explained by the presence of combined hearing and vision impairment and the resulting restricted access to information, their environment and incidental learning opportunities, leading to a heavy reliance on predictability for a sense of security and safety (Probst & Borders, 2017).

Where there is also profound intellectual disability there is also potential for over diagnosis of autism in people with congenital deafblindness (Hoevenaars-van den Boom, Antonissen, Knoors & Vervloed, 2009).

If autism is suspected in someone with CRS, assessment should be carried out by professionals with expertise in congenital deafblindness and with autism (Dammeyer, 2014).

# Care needs

Close medical and dental monitoring is required, even if health seems stable.

Professional that may be involved in assessing, treating and monitoring hearing and vision include

* Ophthalmologist
* Ear Nose and Throat surgeon
* Audiologist
* Orthoptist
* Optometrist and Developmental optometrist (with specialist skills in assessing people with developmental differences).

Other medical specialists may be involved to monitor and treat various body systems

* Cardiologist (for the heart)
* Endocrinologist (for hormonal system)
* Gastrointestinal (for the gut and bowel)
* Rheumatologist & orthopaedic surgeon (for the musculoskeletal system)
* Immunologist (for autoimmune conditions)
* Dentist or dental specialist

Access to an allied health team will assist development and maintenance of well-being, mobility, socialisation and communication. In addition to those mentioned, this team could include a

* Deafblind Consultant
* Orientation and Mobility Instructor
* Speech Pathologist
* Physiotherapist
* Occupational Therapist
* Dietician
* Social Worker

A Psychologist or Clinical Psychologist may also be included in the team if behaviours of concern and/or mental health are limiting a person’s participation and maintenance or development of friendships and connection to other people.

Should the person with CRS have or develop deafblindness a Communication Guide can facilitate day to day communication, interaction, access to information and provide support getting safely from place to place.

During school years the support of a teacher/s with qualifications in special educational need and sensory impairments is also warranted.

# Communication and Independence of people with CRS and deafblindness

There is limited data reporting communication methods and independence of people with deafblindness and CRS. A Canadian study (Canadian Deafblind Association (National), 2014) collected data from 53 people with CRS, aged between 29 and 62 years, the majority of whom had both vision and hearing loss.

Methods of communication included sign language, speech, signed English, individual adapted signs, gestures, body language and objects. Most used a combination of communication methods.

Around 70% resided in a support living arrangement and around 25% lived with their birth family. Approximately 20% were employed in part- or full-time work.

People with CRS and deafblindness are likely to need lifelong medical monitoring and deafblind specialist allied health and care services to enable them to achieve and maintain health and actively participate in home and community life.

There are no known support groups in Australia, specifically for Congenital Rubella syndrome. The [Congenital Rubella Syndrome Facebook group](https://www.facebook.com/groups/congenitalrubellasyndrome/) has an international membership, with some Australian members

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