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# CONGENITAL RUBELLA SYNDROME LATE MANIFESTATIONS: NEW CHALLENGES FOR REHABILITATION AND MENTAL HEALTH PROVIDERS

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## Abstract

Congenital Rubella Syndrome (CRS) is a little known syndrome that is emerging in those people whose mother was infected with rubella while pregnant with them. As the people from the epidemic of the 1960s age physical and mental health issues are emerging, thought to be directly a cause from the rubella virus. Due to the physical and mental health issues these people are showing up in doctor's, mental health professional's and vocational rehabilitation counselor's offices, which have little knowledge of the syndrome. This article is a starting point to learning more about CRS and its effects on the persons affected.

## Introduction

It is important to keep the adage, "The more I know, the more I know that I don't know!" in mind when talking about Congenital Rubella Syndrome (CRS), as the late manifestations of CRS are just now starting to develop and professionals in the medical field do not know the full extent of the impact of CRS. It is also significant to note at this time that not everyone with CRS will develop late manifestations. This information is meant to alert service providers to be aware of this possibility and not to cause alarm.

To understand CRS, it is important to have a clear understanding of the definitions of the various terms used. "Maternal rubella" is the term used when a pregnant woman has contracted the rubella virus. When the developing fetus has been infected with the rubella virus due to maternal rubella, this is called "Congenital Rubella." CRS is a group of symptoms found in about 85% of infants who were exposed to rubella in-utero. The virus may affect all organs by inhibiting cell growth which may cause fetal death, spontaneous abortion, premature delivery or congenital defects.

*Rubella* comes from the Latin word *rubellus*, which means "little red." This term reflects the red rash that develops as part of the illness. It is

also often referred to as the “Three-Day Measles” or “German Measles,” because it was first identified in 1814 as a unique disease in German medical literature. The illness itself is a viral infection that primarily affects the skin and lymph nodes. Historically, epidemics have occurred every six to nine years primarily affecting children and often these children would infect their parent(s). The symptoms can be so mild that the person infected may not know they are ill and as a consequence, about 60% of women who became infected did not realize it. Therefore, if a woman was pregnant at the time and did not realize she had contracted rubella, it would not be recognized that her infant may have CRS. The last major epidemic in the United States was during the mid 1960s, however, there were still some epidemic pockets during the 1970s and 1980s.

When rubella occurs in a pregnant woman, the effects on the developing fetus can be devastating. Children infected with rubella before birth are at risk for growth retardation, mental retardation, deafness and malformations of the heart and eyes in addition to liver, sleep and bone marrow problems. Infants born with CRS can shed the virus in urine and fluid from the nose or throat for a year or more and pass the virus on to others not vaccinated. Since the rubella infection can persist after birth, this suggests that the effects of the virus on brain development may have an on going impact even after birth (Brown, 2001). Alan Brown et al’s 2001 study stated, “...in susceptible individuals, a brain lesion induced by prenatal rubella could initiate a cascade of events that reverberate throughout development.”

A vaccine was developed in the 1960s and first distributed in the United States in 1969. The initial dose is given to children at 12 to 15 months of age and a second dose is given at four to six years old. This vaccine can and does prevent the rubella virus. In the 1990s a debate started regarding the Mumps/Measles/Rubella (MMR) vaccination and whether or not it can cause autism, which resulted in the reluctance of some parents to have their children vaccinated. In March 2005, it was announced that rubella has been “eliminated” in the United States. Due to the disparity in immunization rates from country to country, however, approximately 100,000 babies are still born worldwide with CRS annually. It is important to also keep in mind that 5% of those who receive the rubella vaccine do not develop immunity to the virus.

As stated previously, during the mid-1960s in the United States, there was a major epidemic of rubella. It is estimated that 12.5 million individuals

were infected with rubella during this epidemic. Of the 12.5 million, it is estimated that 20,000 children were born with CRS and of the children born with CRS, there were more than 11,000 born deaf, 3,580 born blind and 1,800 born mentally challenged. Many of the mentally challenged children were also both deaf and blind. In addition, more than 2,000 infected children died in the first four weeks of life. As these children with CRS aged, additional late onset medical problems began to emerge.

The most frequently identified “early manifestations,” those evident between birth and adolescence, are hearing loss (73%) and visual impairments. The most common visual impairments are cataracts (20-50%) and microphthalmia, meaning undersized eyes (50%). Other early manifestations include congenital cardiac problems, mental retardation (42%), and autistic-like behavior (7.4%). There may also be neurological problems including microcephaly (abnormal smallness of the head), cerebral palsy, hypotonia (decrease of normal tonicity or tension as in muscles or arteries), poor balance and dyscoordination (impairment of the ability to perform smoothly coordinated voluntary movements). Most of these manifestations are apparent at birth; however, some may emerge two to seven years after the original rubella infection even in the children who had no symptoms at birth. Two of the most common of these early delayed manifestations that were not present at birth are hearing loss and vision loss, (Polowe-Aldersley, 1990).

Now, 30 to 40 years after the last major epidemic, we are seeing the emergence of “late manifestations” of CRS which may start in the adolescence years or later. Since this is fairly new information, it is unknown if the late manifestations will end at some point or if they will continue to develop. Some of the late manifestations being observed in individuals who are deaf from CRS are a change in hearing ability; visual problems (most common are acquired glaucoma and detached retina), diabetes, thyroid conditions, gastrointestinal difficulties, vascular problems, seizures, effects to the central nervous system, and early menopause. In some cases there are also psychological problems that may emerge including withdrawal, reduced frustration threshold, depressed mood, and emerging psychotic symptoms. Behavioral problems are also becoming apparent with the most common being impulsive behavior and attention deficiencies.

There are some psychiatric and neurological symptoms associated with CRS that need a closer look especially the appearance of a 20.4%

risk of adult onset of schizophrenia or schizophrenia spectrum disorders. Research has shown that these psychological problems are not related to the individual's deafness because they also appear in CRS adults that are not deaf. In research studies it was found that rubella exposed infants are at a greater risk of developing schizophrenia when they become adults. In one study, MRI tests show people who have CRS and schizophrenia like symptoms had reduced cortical gray matter volume and greater size of the lateral ventricles, finding that have been shown in studies of persons with just schizophrenia, (Brown, 2001). In addition, research shows that there is a decline in IQ from childhood to adolescence (Brown, 2001).

A major question at this time is regarding the possible causes of these late onset conditions and there are several prevailing theories. One theory is that the virus persists in the affected organ(s) or that the old infection sets up an autoimmune response (Nicholas, 2000). Another theory is that due to some cause or circumstance, the virus reactivates. A virus is not a living organism and cannot be killed but can remain dormant for many years then reactivate causing additional problems. The most popular theory at this time is that the effects of the original virus were so devastating that the organs and systems start to deteriorate more quickly as the individual ages.

Much research needs to be done to address all these various issues, especially regarding whether or not prenatal rubella exposure is a cause for schizophrenia and/or autism. It is hoped that a major research project will soon be undertaken focused on studying the late onset manifestations of CRS in individuals who are deaf. In the interim, Nancy O'Donnell at the Helen Keller National Center has developed a data collection form that can be used in collecting preliminary data for future research. It appears that what we know so far is only a small portion of the larger picture.

Meanwhile, the implications for rehabilitation, mental health and physical health professionals are vast. This issue first came to the attention of vocational rehabilitation counselors for the deaf in Kentucky when consumers started returning for services following perhaps years of successful employment. Sometimes dramatic changes were noted that had occurred since they last passed through the system. Most notably were the myriad of mental health issues that needed addressing before beginning preparation for re-employment. Often there were also medical problems (primarily diabetes, thyroid and heart problems) that must be addressed and considered

in job placement. Behaviors, including impulsivity, low frustration tolerance, aggression, mood shifts, compulsive behavior and decreased attention span often necessitated a referral for supported employment in lieu of returning to competitive employment. The issue then arose that due to the years of successful employment and stability the consumer may not meet the qualifying criteria for supported employment. Also, referrals needed to be made for a visual evaluation due to the increased possibility of development of cataracts and glaucoma and a thorough physical evaluation for possible adult-onset diabetes, heart problems, thyroid problems and respiratory problems, all of which can impact job placement and job continuation.

Implications for the mental health professional are similar to that of the rehabilitation professional. Many of the physical problems that are emerging can have an immense impact on a person's mental health status. For example persons with untreated thyroid conditions, heart conditions and diabetes can have mood swings or depression specifically due to the illness. Then add in the feeling that the person is "falling apart" and can not do things they used to be able to do and this certainly impacts a person's mental health. There have also been some atypical symptoms such as a late onset of psychotic symptoms. In the general population the onset is often in the persons late teens or during their 20s. Persons with CRS are starting to experience some psychotic symptoms in their late 30s and into their 40s and their presentation are somewhat different. From observation of clients with psychotic symptoms and possible CRS the negative symptoms of schizophrenia are minimal and they respond well to medication. In some cases, a decline in cognitive abilities has been noted. Clients known to have CRS often report "forgetting" things, such as appointments or conversations they have had as well as previously learned skills. As a consequence, when working with individuals with late manifestations of CRS it is important to educate them on how to cope with their various long term illnesses as well as teaching them new skills to help them adjust to their memory loss.

The medical community is, for the most part, unaware of late manifestations of CRS. Due to the diversity of symptoms, the individual is often referred to a variety of doctors with expertise in one or the other particular area. As a consequence, there is no centralization of the symptoms to enable one to recognize they may all be related to a particular syndrome. Therefore, it may be necessary for the vocational rehabilitation or mental health professional to advocate and educate for the client.

The bottom line is that we are all still learning about CRS and recognize that there is still much to be learned. Due to the effects of these late manifestations on the life of individuals affected, including employment, physical and mental health, the need becomes urgent for more research and information. At this point, the more this is discussed, the more the word is spread and the more information is shared, the more we will learn and be able to better serve those who are experiencing late manifestations of CRS.

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