Congenital rubella syndrome: 30 year after the epidemic of the 1960's.

Authors:
O'Donnell, Nancy

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Abstract:
Describes the efforts of the Helen Keller National Center to gather information about the incidence of late onset medical and behavioral problems from parents with children who are deaf-blind as a result of congenital rubella syndrome in the United States. History of the disease; Description of symptoms.

CONGENITAL RUBELLA SYNDROME: 30 YEARS AFTER THE EPIDEMIC OF THE 1960'S

Contents

This article describes the efforts of the Helen Keller National Center to gather information about the incidence of late onset medical and behavioral problems from parents with children who are deaf-blind as a result of congenital rubella syndrome (CRS). A national survey was conducted from 1990-1991, tapping into parents as the main sources of information about their children. The results indicated that many individuals with CRS are not exhibiting additional problems. However, of those who do describe symptoms, they range in severity from late onset glaucoma, diabetes and/or thyroid problems to erratic changes in behavior in individuals with degenerative processes.

History of the Problem

Rubella, also known as German measles, is not usually considered to be a serious childhood disease. Its symptoms, including rash, runny eyes, swollen glands, low-grade

fever, pain in the joints, and sore throat, are sometimes so mild that some individuals never even realize that they have been infected. Serious problems can occur, however, when a woman contracts rubella in her first trimester of pregnancy. During this critical period in fetal development, the virus can infect and affect virtually every developing organ.

in the early to mid-1960's, an epidemic of rubella spread across the United States, infecting approximately 1 out of every 10 pregnant women (Vernon, et al., 1980). As a result of maternal rubella, an estimated 20,000 children were born with a variety of handicapping conditions including hearing loss, cardiac problems, vision loss, mental retardation, and other developmental disabilities (Preblud, et al., 1980). This constellation of symptoms came to be known as congenital rubella syndrome, or CRS. Trybus et al. (1980) reported that 37 percent of children who were reported to be deaf as a result of maternal rubella had one or more additional handicapping conditions. Estimates on the number of children who were born with deaf-blindness varied from a low of 739 to a high of 10,000 (Lockett & Rudolph, 1980).

During the mid- to late 1980's, the Helen Keller National Center (HKNC) began receiving calls for help from workers and parents around the country who were working and living with persons who were deaf-blind due to CRS. These callers described sudden and inexplicable changes in the behaviors of these individuals, including disorientation, decreased attention span, and increased or decreased activity levels, as well as self-abuse and aggressive behaviors.

To address these concerns, HKNC turned to staff at the Developmental Disabilities Center of St. Luke's-Roosevelt Hospital in New York City who had been working with individuals with CRS since the epidemic of the 1960's. They suggested that these behaviors could be tied into late onset manifestations of CRS. Some of the physiological changes which they had observed in patients in their clinic were higher than expected incidences of glaucoma, diabetes, and thyroid dysfunction. Because there had been very little recent information written about this phenomena, HKNC decided to try and gather further information through its own survey.

Procedure

The survey was conducted in two phases. In Phase I, which took place from November 1989 to April 1990, a staff person interviewed parents/caretakers by telephone and filled out a survey form during these conversations. In Phase II--June 1990 to April 1991--a large scale mailing was conducted across the country. In both phases, we sought out parents as our primary sources of information. We felt they would be the most reliable sources of long-term medical information as well as anecdotal information.

Information was requested about each person's status in the following areas: current residential placement, current employment/day placement, hearing, vision, communication methodology, opportunities to communicate at home and work, increases or decreases in various behaviors over the past year, life changes experienced over the past year, medical conditions, age of onset, and medications.

During Phase I of our effort, 39 interviews from parents in 19 states were completed. Much of the information received during these interviews was anecdotal in nature. Based on this feedback, we revised our survey form. We then determined that Phase II should be conducted by mail to reach a larger number of potential respondents.

During Phase II, we received and analyzed 88 responses representing another 14 states. The percentages reported herein are based on those 88 responses. However, the impressions, quotations and interpretation of the data also reflect the wealth of information gathered from the initial 39 interviews.

The following report is divided into two main sections. Section I provides a general overview of the respondents. Section II focuses on medical information.

I. General Overview of Respondents

The survey targeted persons aged 16 and older reported to be deaf-blind as a result of CRS. Fifty-eight percent of the responses were in regard to males, 42 percent were females. The largest percentage of respondents (32 percent) fell into the 25-26-year-old category, reflecting individuals who were born as a result of the rubella epidemic of the 1960's.

Residential Profile. Forty-one percent were reported to live with parents/family, almost 20 percent were in some type of group home, 11 percent lived in MH/MR facilities (institutions, state hospitals), 9 percent were in foster care, and 9 percent were in an intermediate care facility.

Employment Profile. Approximately 32 percent worked in a sheltered workshop or work activity center; 27 percent were students; 20 percent were in a training facility, including employment training; 2 percent were competitively employed; and 3 percent were engaged in supported employment.

Hearing and Vision. Almost 50 percent reported little or no useable vision. Thirty percent reported significant additional problems.

Approximately 90 percent were considered to have no functional hearing, 45 percent fell into the category of profoundly deaf with no vision, and another 28 percent were profoundly deaf with significant vision problems:

Communication. The following percentages indicate a yes/no response to a particular method of communication. It does not rate the capability of the individual in each category. Respondents were allowed to check as many methods as applied.

In terms of receptive methods, out of a list of 10 possible methods for receiving communication, sign language was the most frequently indicated, with 83 percent. Gestures and prompting were checked by 60 percent of the respondents. One fourth of the cases were reported to understand fingerspelling. Five individuals, or 6 percent, could reportedly read braille, and 3 individuals, or 3 percent, were described as having no receptive method of communication.

With a 70 percent response, sign language was the most common method of expressive communication; 55 percent were reported to use gestures; 11 percent used speech, either alone or in combination with another method; and 6 cases, or 7 percent, reported no method of expressive communication.

Opportunities to Communicate. When asked if there were someone in the deaf-blind person's living situation who can communicate effectively with him or her, 16 percent of the respondents answered "no." Twenty-three percent answered "no" when asked the same question in response to the work placement/day program.

Behaviors. Respondents were asked to indicate which out of nine behaviors were currently demonstrated by the deaf-blind person and whether the frequency of these behaviors had increased, decreased, or remained the same over the past year. Self-stimulation and vocalization were the most commonly occurring behaviors, often reported every day. Approximately 60 cases, or 68 percent, reported that this behavior decreased over the past year. Disruptive activities were the next most frequently noted behaviors. Ten people, or 11 percent, were reported to do t his every day, whereas 40 percent were disruptive only occasionally. Approximately one-third of the respondents were not disruptive at all. Tantrums/outbursts were noted at least once a week in 35 percent of the cases. This behavior occurred occasionally in one-third of the individuals. Almost 20 percent did not demonstrate this behavior at all. The frequency of this behavior was noted to have decreased or remained the same in 60 cases, or 68 percent, over the past year. Self-injurious behavior was reported to occur at least once a week in 3.5 percent of the cases. Forty percent did not demonstrate this behavior. About 40 cases, or 45 percent, indicated a decrease in this behavior since the past year.

The most serious demonstration of behavior problems, damages to property and assaults on others, were reported to occur in 11 cases (12.5 percent) at least once a week. Thirty-four percent were assaultive occasionally, while 28 percent damaged property occasionally. Approximately 50 percent did not demonstrate either behavior. In general, there was a definite trend of all behavioral categories remaining the same or decreasing over the past year.

Lifestyle Changes. Individuals with CRS have sometimes been described as having difficulty accepting changes in their routine/environment. Information was requested on the number and types of changes that occurred in each person's life during the past year. The results were as follows: Twenty-seven percent experienced a change in health, 26 percent had a change in living arrangements, 43 percent had a change in family/ roommate/staff (residential), 33 percent had a change inwork, 36 percent had a change in staff/fellow employees, and 18 percent had a change in transportation.

Many people experienced changes in more than one category and several had changes in all categories over the past year. In considering reasons for sudden and inexplicable changes in a person's behavior, these factors should be taken into consideration.

II. Medical Conditions

This section is divided into seven categories: auditory disorders, ocular damage, cardiac problems, endocrine system (diabetes and thyroid), degenerative conditions, and esophageal/gastrointestinal symptoms.

Auditory Disorders. Several authors have addressed the issue of hearing loss associated with CRS. Vernon et al. (1980) indicated that it was the most common handicap associated with rubella, affecting about 73 percent of the cases. They described these losses as sensorineural in nature and generally bilateral. Among these, Waxham and Wolinsky (1984) stated that by 7 to 9 years of age, nearly 93 percent had some auditory problem. Sever et al. (1985) talked about some cases in which children with CRS whose audiograms and speech patterns had been normal suddenly developed mild to profound sensorineural hearing loss. The latest age at which they found documentation for this occurrence was 10.

Ninety-two percent of our respondents in Phase II indicated that their child had little or no hearing at the time of the survey. One case of late onset hearing loss was reported to have occurred at age 10.

Ocular Damage. In 1980, Vernon et al. reported that approximately 33 percent of rubella youths have visual defects. Sever et al. (1985) cited one study in which almost 10 percent of patients with prior ocular damage developed additional forms of eye damage as a delayed manifestation.

According to Sever et al., cataracts occurred in 25 percent of those infected between 25-93 days gestation. Vernon et al. (1980) indicated a 20-50 percent occurrence of cataracts in rubella infants.

Sixty-six percent of our respondents were reported to have cataracts, with 57 percent indicating that these were congenital. It should be expected that our percentages would be higher than those in the literature because we exclusively sought out individuals with vision and heating problems rather than surveying the whole population with handicapping conditions from CRS. Two cases of cataracts were identified at age 7, one case at age 16 and one at age 18.

Seaver et al. cited Boger's report of glaucoma with late onset in 13 patients with CRS. This diagnosis was first made when the patients were between 2 and 22 years old. In 11 cases, the eyes were microphthalmic, two had eyes of normal size. All had cataractous lenses either removed surgically or absorbed spontaneously. Vernon, Grieve and Shaver (1980) reported that glaucoma was present in 4 percent of rubella infants.

Thirty percent of our respondents were reported to have glaucoma, 7 percent of which had been diagnosed at age 1 or younger. This rate of occurrence is very high, when compared to an incidence of 0.5 percent of glaucoma in the general population, Although glaucoma can develop as a poor outcome of cataract surgery, Fedun (unpublished observations, 1991) noted that late onset glaucoma has occurred in children who never had cataracts and in those whose unilateral cataract was never operated on. Her observations include those who are deaf due to CRS with no previous vision problems

who have been found to be developing glaucoma at much higher rates than the general population.

When we asked survey participants if glaucoma testing had been performed in the past year, only 32 percent responded in the affirmative. We strongly recommend that glaucoma testing become part of the annual physical examination. Glaucoma prevention is critical in order to try to maintain whatever vision an individual has, particularly in light of additional handicapping conditions. We have had reports of glaucoma that is difficult to control by medication and cases where the eye had to be surgically removed (enucleated).

We could find no documentation describing an expected rate for detached retinas in the CRS population. Our survey showed 12 cases of individuals with detached retinas (13 percent). For those who reported an age of onset, there was one case diagnosed at each of the following ages: 2, 8, 10, 12, 16, 17, 18, 19, and 39. Thirty-five percent of our cases were reported to engage in self-injurious behavior at least once a week. Some of this behavior (head banging, eye poking) could undoubtedly contribute to retinal detachment.

Cardiac Problems. Heart problems were fairly common in the CRS population. Vernon et al. (1980) cited statistics of 35-76 percent of rubella youth with heart disorders, including up to 58 percent with patent ductus and up to 18 percent with ventricular septal defect. Waxham and Wolinsky (1984) reported that heart defects occurred in 30 percent of children infected between 25 to 93 days of gestation. In terms of late onset cardiac problems, we can look to a longitudinal study which was conducted on 53 Australian subjects who were born with CRS between 1939 and 1943. McIntosh and Menser (1992) reported that, as of 1991, 3 of the 50 subjects had died of cardiovascular causes.

Fifty-two percent of our respondents indicated the presence of a heart condition. Patent ductus, atrial, or ventricular septal defects and pulmonary stenosis were the most common. In many situations, surgical intervention had occurred in infancy. Twenty-eight out of 88 participants, or 32 percent, answered affirmatively when asked whether or not cardiac testing had been done in the past year. In 1990, one of our survey participants died from congestive heart failure at the age of 25.

One mother described a large hospital's ongoing study of congenital heart defects. As part of the research, her daughter was put on a Halston heart monitor for a 24-hour period. Surprisingly it was found that her heart stopped beating several times, missing up to 5 beats at a time. In 1987, at the age of 24, she received a pacemaker. In February 1990, when her mother was interviewed, all was well. At the beginning of 1991, the mother called to report that her daughter had since developed tachycardia (an abnormally rapid heart rate tachycardia). She was also retaining fluid in her feet and she was put on medication for both of these conditions. A previous occasional problem with gagging had recently worsened to the point where it was occurring on a daily basis. No explanation is known for the development of these conditions.

Endocrine Systems. Forrest et al. (1971) reported on a followup of 50 congenital rubella patients in New South Wales, Australia. Five out of the 50 patients had developed

diabetes mellitus and 4 had latent diabetes by the time they were 35 years old. Mcintosh and Menser (1992) did a 50-year followup of the same group and found that these rates had remained the same.

Our survey indicated that 5.7 percent (5 out of 88) of our participants were diabetic. The ages of onset were reported to be 2, 5, 14 and 24. Although our percentage is lower than those indicated in previous studies, the bulk of our survey participants were 25-26 years old or younger, as opposed to those in the Australian survey, who were approximately 35 years of age. It remains to be seen what the next 10 years will show us in our population. Sever et al. (1985) commented that the high prevalence of diabetes in patients with CRS is remarkable, since diabetes mellitus develops in the general population in only approximately 0.1 percent of individuals younger than 30 years of age.

Please note that several parents also mentioned that their child's blood sugars fluctuated a great deal. This has been observed by Fedun as well in her ongoing study of the relationship of diabetes and CRS (Fedun, unpublished observations, 1991). Efforts are continuing to obtain more information about this phenomenon.

Regarding thyroid disease, Sever et al. (1985) stated, "Hypothyroidism, hyperthyroidism and thyroiditis have all been reported as delayed manifestations of CRS." They cited one study which identified thyroid dysfunction in 5 percent of patients with CRS. An autoimmune response has been indicated as a possible cause of thyroid disease. Our survey reported 6.8 percent, or 6 out of 88 respondents, with thyroid problems. Onset, when reported, occurred at 11, 20, 26 and 35 years of age.

Degenerative Conditions. During the course of this survey, several parents and professionals have raised questions regarding degenerative processes which they have observed in some individuals. Often, the discussion would turn to a condition called progressive rubella panencephalitis (PRP). First recognized in 1975, PRP is a very rare degenerative process of which there were only 12 known cases in 1984 (Wolinsky, 1984). It has been documented in individuals with CRS as well as those with naturally acquired rubella. All of the cases were male. The literature outlined the following symptoms of PRP, which were typically first noted between the ages of 8 and 19: Deterioration in school performance and behavior, sometimes accompanied by seizures, may be observed first. The gait is noticed to become clumsy early on and, in time, a progressive global dementia follows. Ataxia (muscular incoordination) progresses to involve all limbs. The degenerative process can reportedly continue for many years. Wolinsky and Slagle (1989) reported that all known cases of PRP were fatal. However, Wolinsky and McCarthy (1991) also cited Martin et al. (1989), who suggested that spontaneous remission of symptoms appears possible.

The following survey excerpts will provide a glimpse into the types of degenerative symptoms that some of deaf-blind adults with CRS are exhibiting. In all, we had five cases describing noticeable and sometimes severe degenerative processes. Some of our cases differ from the above description of PRP in that the first symptoms were noted when the person was in the third decade of life. Also, two of the five cases were females.

We are not presenting these cases as examples of PRP, but rather as examples of individuals with degenerative processes.

The first case describes a 26-year-old male: "Over the past 4 years, [our son] has gradually lost his ability to walk. In the past year, he cannot stand by himself nor walk at all without totally leaning on another. . . . [He] was always very hyper. . . also very aggressive. We were recently told that [he] is on a downhill slide and will eventually lose all control, including muscles, swallowing, etc. . . . The doctor does not know another patient with [this] condition. The doctor advised us to remodel our house to accommodate a wheelchair. Do you know how long a time between wheelchair and bed?"

Another parent gave this description of her 24-year-old daughter: ". . .about 6 years ago [she was 18] things first changed. She had one seizure. She became much more irritable, broke lots of windows, would actually swing at people close to her, wasn't as willing to do things she had always loved, and was generally terrible. Her tantrums were frequent, violent, and longlasting. This went on for 2 to 4 years to some degree. About 2 years ago she calmed down some, but in November 1988 all at once she became very unsteady on her feet. I thought she may have had a stroke. Her gait is much like that of an individual with cerebral palsy. She looked 'drunk' all the time. She had a great deal of trouble walking 3 feet without hanging onto something. She appeared real confused over this change of events and began to really depend on us to help her get from place to place. But the tantrums stopped. At this point we had a CAT scan done, took her off her phenobarb, took her to a heart specialist, otorhinolaryngologist, neurologist, and just about everyone else we could think of--no one knows what's going on. Recently, I requested an MRI. That did show a large 'scrambled-egg'-appearing thing. The report ruled out a brain tumor, mentioned in passing, and rejected multiple sclerosis, so now we know there is something in/on her brain but don't know what it means. They also don't know if it is something new or has always been there. How I wish we had been able to have tests like this when she was a baby. Right now she has been 'off balance' for a year and a half. She has adapted her once springy gait to a wide stance, is sometimes amused, sometimes frustrated, over her new limitations (above I observe from behavior and facial expressions--she can't tell me these things) and does not seem to have regressed any more during the last year and a half."

Correspondence from this mother in April 1992 updated her daughter's condition. "Her late onset behaviors have changed again. The temper tantrums have diminished, but now she staggers and falls easily."

Esophageal/Gastrointestinal Conditions. We first heard about problems with swallowing, gagging, and vomiting during conversations with several parents who participated in the telephone survey. You may recall the young woman described in the section on cardiac problems who also developed a problem with gagging. Phase II of the survey brought us more examples of this behavior.

One parent described these symptoms in her 19-year-old daughter: "The past year [especially 3-5 months], we have had some strange behaviors come and go with [our

daughter]. [She] is definitely experiencing changes, whether physical or emotional is anyone's guess. She has had some weight loss, decrease in appetite with intermittent crying spells at times; when crying, she does a strange throat sound prior to this--then just sobs. During these times she gags violently as if to vomit but doesn't. If anything, it's only mucous. She has had most exams done blood tests, urine, complete dental checks under anesthesia--and we have found no obvious physical ailment."

This was written by the mother of a 24-year-old daughter: "The doctors are unable to determine the cause of her recurring vomiting (every 3-9 weeks for past 6 years)."

We have been unable to find any mention in the literature about these symptoms. However, since we first reported on this problem, other cases have been brought to our attention. Interestingly, we have been contacted by others outside the United States who have also observed this phenomena: a mother of a 16-year-old daughter in England, another mother of a 5-year-old daughter in England, and recently a mother of a 14-year-old girl in Canada. In the summer of 1991, we received a letter from a doctor in Sweden who cited 3 patients with CRS who had gagging and vomiting problems, with no apparent medical explanations. We will continue to collect data about this phenomena and would appreciate any insights which the readers may have.

In addition to the conditions described above, parents have noted other changes. These include: seizures, reported in about 10 percent of the cases; large weight gain, especially in women; hirsuitism (excessive hair growth) in women, especially on the face, possibly on the chest; premature aging; psychological/psychiatric problems; and loss of bowel/bladder control, sometimes on a temporary basis.

General Conclusions

When we started this project, we did not anticipate the scope or severity of some of the medical problems this group is experiencing. Although almost a third of our respondents indicated no late emerging medical problems, the other two-thirds reported problems which ranged in severity from individuals who only developed glaucoma to one case of a woman who developed every late onset symptom on our list except diabetes. Researchers still do not understand the mechanism which triggers the development of these conditions, the probability of any one person developing a particular condition, or a timeframe in which we might expect these things to occur.

in looking to the most recent data from Australia's 50-year followup of 50 patients with CRS (McIntosh and Menser, 1992), we are unable to glean much insight into what the future might hold for our population. Theirs was a group which, in general, was less affected than ours.

If we look at our survey respondents as a whole, we see a severely involved group of men and women, many profoundly deaf and totally blind, or with significant vision problems, many with limited communication systems, some with limited opportunities to communicate. They are undergoing, or have the potential to experience, significant changes in their medical condition, yet are very limited in their ways to tell us about their

symptoms. They have often been subject to changes in their living and/or working situation, yet have probably been unprepared for these changes because of their own limited communication or that of the people around them. If we feel any sense of urgency in relation to this population, it is, of course, in regard to their medical status first. However, one simply cannot ignore the critical area of communication.

For more information, contact: Nancy O'Donnell, Helen Keller National Center, 111 Middle Neck Road, Sands Point, New York 11050-1299, (516) 944-8900. (TTY and Voice)

PHOTO (BLACK & WHITE): Learning to use the Telebraille.

PHOTO (BLACK & WHITE): Using tactile sign language, interpreters talk with persons who are deaf-blind.

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By Nancy O'Donnell

Ms. O'Donnell is Special Project Coordinator for the Helen Keller National Center.

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