Health, well-being and congenital rubella syndrome

A Sense and Sense Scotland practice sharing event
6-7 March 2009, Crewe, England

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Introduction
Sense and Sense Scotland were founded because of rubella. When pregnant women catch rubella their babies are affected by the virus and many are born deafblind and with other difficulties. Fifty years ago families desperately needed support and came together to find it. Over the years, we have built up considerable knowledge and experience together and this seminar enabled us to share our knowledge of congenital rubella syndrome, and to learn from each other and from colleagues in other organisations and in other countries.

In the UK today, few babies are born with rubella thanks to the almost universal vaccination of our population through the MMR programme. But people are still living with CRS – they are getting older now, and we need to continue to research and learn about the combined effects of age and CRS on people’s lives.

Elsewhere in the world too, rubella is still a major problem and there will long be a need for the knowledge we are gathering now.

During the seminar over 50 delegates – most of them practitioners – talked about what it’s like to grow older with congenital rubella syndrome, and what we can do to support people better. While there was plenty of information and discussion about the medical health aspects of CRS, the dominant desire of everyone present was to find ways of ensuring that people with CRS experience the best possible quality of life, no matter what health issues they live with.

A trio of overarching themes developed:

- the importance of understanding the initial and ongoing medical effects of CRS and their implications
• the importance of communication between professionals and the person with CRS

• the importance of collaboration between professionals.

This report describes the main ideas and information discussed at the seminar. Each section begins with a brief summary so that you can skim through the report quickly – or you can read the more detailed information which follows.

The report follows the order of the presentations and workshops over the two days of the seminar. The first day focussed primarily on medical health issues, and the second on people’s well-being and the effects of CRS on behaviour.

The discussions centred on the lives of people severely affected by CRS, many of whom have serious multiple impairments and communication problems, although we recognised that CRS affects people in different degrees and that people less seriously affected also want to know the long-term health effects of the syndrome.
1 An overview of health and congenital rubella

Linda Long, Health Development Officer, Sense Scotland
Pat Tookey, National Congenital Rubella Surveillance Programme, UCL Institute of Child Health, London

Summary
Rubella is a mild illness, mostly caught in childhood. However, when transmitted to a woman in the first four months of pregnancy it can cause serious impairments in the baby, leading to eye, ear and heart defects and other problems.

In much of Europe and the Americas rubella is now almost absent because of comprehensive immunisation programmes, though elsewhere rubella is still a significant problem. Even where new cases of CRS are rare, adults continue to live with CRS: the purpose of this seminar is to look at their lives, and how they are affected by CRS.

Though scientific research into the effects of CRS on adults is still limited, we do know that people with CRS are more likely to suffer thyroid conditions and diabetes than other people. We also suspect that CRS may cause other conditions to develop earlier or more often.

Sharing your knowledge
There are two groups online where you can share and learn more about CRS. Both are moderated by Nancy O’Donnell at the Helen Keller Centre in New York. One group is for people affected by CRS (many were at school together in Boston, USA): it’s busy, and has two key professional contributors, Nancy O’Donnell and Pam Ryan from Perkins school. The other group is for worldwide professionals working with deafblind people. To find out more about joining, email Nancy O’Donnell at HKNCNOD@aol.com.
Finding out more
Sense’s website: www.sense.org.uk

DBI rubella network:
www.deafblindinternational.org/standard/network_rubella.html


NCRSP monitoring:
http://www.ich.ucl.ac.uk/ich/academicunits/Paediatric_Epidemiology_and_Biostatistics/Research

2 Endocrine problems

Chris Fuggle and Gini Bartlett, Sense

(with support from Thyroid UK and Diabetes UK)

Summary 2
The way that hormones work in our bodies is extremely complex and not yet fully understood. We do know, however, that people with CRS are more likely to develop certain hormone-related problems, in particular thyroid problems and diabetes.

The thyroid gland may become over-stimulated and over-produce thyroid hormones (hyperthyroidism), which essentially causes the body’s systems to run at high speed. Alternatively, if the thyroid is underactive (hypothyroidism) the body’s systems slow down. Both conditions can be treated with medication.
Diabetes is extremely common, and is caused by a lack of insulin or the insulin in the body not working effectively. This means that the body cannot absorb glucose efficiently, and so cannot derive energy from food appropriately. Though a small proportion of diabetics need to inject insulin, most can control the problem by managing their diet, exercise and lifestyle.

More information on diabetes and thyroid problems
You can find a great deal of useful information and support from specialist organisations:

Diabetes UK
Macleod House,
10 Parkway, London NW1 7AA
Tel 020 7424 1000
Fax 020 7424 1001
Email info@diabetes.org.uk (if you use aol, Diabetes UK recommends that you write or use the phone)
Web www.diabetes.org.uk

Thyroid UK
32 Darcy Road
St Osyth
Clacton On Sea
Essex
CO16 8QF
Tel 01255 820407 (permanent answerphone)
Web www.thyroiduk.org
3 Vision

Lucy Butler, consultant ophthalmologist
Mary Foster, Sense West multi-sensory impairment advisor

Lucy and Mary work closely together in Birmingham  Lucy talked about the eye conditions that people with CRS have. Mary contributed case studies and suggestions for practitioners.

Summary 3

A defining feature of CRS is the eye problems that people are born with. For example, many babies are born with cataracts – which nowadays are operated on, and replacement lenses inserted. Some babies may be born with small eye and most have some vision problems.

As people with CRS grow older it seems that they are more prone to other eye conditions than the general population. The most common conditions in people with CRS are cataracts, glaucoma and retinal detachment. In addition, because more people with CRS develop diabetes, they are more likely to develop diabetic retinopathy.

It is therefore crucial that everyone with CRS has regular eye checks. For some people these eye checks can be difficult – it is not unusual for people to be uncomfortable in a clinical setting, and all eye tests require some degree of co-operation – but it is worthwhile for everyone. The key to successful eye checks is to build good relations with the eye clinic and to prepare well.

It is also important to be aware of the signs of deteriorating vision – such as a person becoming clumsier, or finding it hard to judge the height of steps and curbs. Staff need to watch out for these symptoms and to seek help urgently as they can indicate a serious problem that needs immediate treatment.
Feedback from delegates
Many delegates were interested in sharing ways of preparing people for medical exams – this information was very useful.

There was also much discussion about how to collaborate more effectively with other professionals. The lack of time during an appointment was a real challenge.

Communication passports are very useful to save endless repetition when dealing with healthcare professionals.

Further information on vision
The RNIB has an excellent range of factsheets and leaflets, and more advice on its website. It’s worth contacting its Multiple Disability Service at

224 Great Portland Street
London W1W 5AA
Tel 020 7388 1266
www.rnib.org.uk and www.rnib.org.uk/multdis

Useful RNIB Focus factsheets are:
Looking for eye problems in people with learning difficulties
How to get the best out of sight testing for people with learning difficulties
Questions to ask the Optometrist after the eye test of an adult with learning difficulties
Obtaining services for people who have sight problems and additional needs
Making sense of prescriptions for spectacles
Minimising problems in eye surgery for adults with severe learning disabilities

4 Hearing
Jenny Fletcher, Sense West Deafblind Consultant

Summary 4
Ear problems are the most common effect of CRS and are present from birth. Because both the inner and middle ear may be damaged this may cause problems not only with hearing but also with balance and movement. As people get older they may also experience age-related hearing loss and loss caused by damage to the ears by loud or persistent noise.

The most common ear problems experienced by people with CRS are tinnitus, recruitment (where certain frequencies are very loud), loss of balance, auditory processing disorder, and problems with integrating the senses.

It is often hard to tell the effects of damage to the ear as clinical tests are at best a rough guide. We therefore also carry out extensive functional tests to see how someone hears, and how different situations affect this.

Feedback from delegates
One delegate commented ‘I learned more about hearing issues than I ever thought about before.’

Many delegates found the information on the possible effects of tinnitus and recruitment on behaviour particularly interesting.
5 Heart conditions
Linda Long, Sense Scotland

Summary 5
This workshop looked at the type of heart problems that people with CRC may experience, and also provided advice about how to support a deafblind person who is receiving treatment in hospital. The most common difficulties are:

patent ductus arteriosus (PDA): the child has a hole in the heart that has to be closed by surgery or some type of plug.

pulmonary artery stenosis: this is where there a narrowing of an artery causes reduced blood flow to the lungs and puts a strain on the heart. The artery can be widened by inflating a balloon via a catheter.

As well as this medical information, there were a number of useful discussions about how to support a deafblind person who is receiving treatment in hospital – such as familiarising them with the room where they will be staying.

Feedback from delegates
Delegates were keen to have more information about circulatory problems. They were also clear about the importance of improving communication with medical doctors, and sharing strategies for doing this.

Handy heart sites
www.mayoclinic.com/health/congenital-heart-defects/CC00026

http://kidshealth.org/kid/htbw/heart.html
It’s also worth looking at the British Heart Foundation’s information – they have some excellent leaflets.

6 Healthy living and the role of health detectives
Linda Long, Health Development Officer and Paul Hart, Principal Officer, Sense Scotland

Summary 6
This wide-ranging session moved away from the medical focus of the previous sessions to reinforce the importance of living healthily: making friends, keeping active, eating and drinking well and having a sense of purpose.

‘Health is a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity.’ (WHO, 1948)

Communication is at the root of healthy living – without it we cannot form relationships, make friends, or carry out meaningful tasks.

By enabling people to take part in activities and to meet others while doing so we can help them to build relationships, develop their communication skills, and to find a sense of self through meaningful activities. When everyone – service users and staff - enjoys activities and those activities are memorable people build stronger relationships.
The session emphasised the crucial role that staff play in acting as health detectives – observing and recording what they see, and reflecting on it to find out what health issues people may have, and what we can do to help them. It also gave tips on how to be taken seriously when working with health professionals.

**Conclusion – the key issues**
The key areas where we can help people to live healthy lives are:

- communication – people with CRS, researchers, practitioners and families all need to talk to each other
- relationships – between individuals, organisations and countries
- meaningful experiences.

Bob Snow rounded off the session and the first day by commenting that the seminar had been a meaningful experience for him because it had ‘been like the old days – talking about rubella, communication etc. No one’s talked about personalisation or budgets. It’s been wonderful!’
7 The brain and relating to the world: CRS and behaviour
Dr Jude Nicholas, specialist in clinical neuropsychology is currently attached to the Regional resource Centre for Deafblindness and the University Hospital in Bergen, Norway

Summary 7
We all know that the stage of foetal development at which someone is affected by rubella determines the nature of the damage to their eyes. What many of us didn’t know was that the same applies to the brain.

The rubella virus can damage the effective development of the neural paths in the:

- sensory systems
- motor systems
- cognitive and integrative systems.

These systems develop from gestation into adulthood, starting with the sensory systems and finishing with the cognitive and integrative systems. Early damage due to rubella may not have observable effects straight away – we may only see the effects when the affected part of the brain is called into action as the person develops.

While we can use standard checklists to assess a person with CRS’s behaviour, looking for possible neural impairment, we must always bear in mind the powerful effect of deafblindness itself. So while a person’s changing behaviour may be due to impairments in the brain, it may also be due to the way that deafblindness affects their ability to communicate and understanding of the world. So perhaps we should see abnormal behaviour as normal behaviour which we haven’t yet understood.
There is much research to be done in this field and scientists and practitioners need to work closely together.

**Sources and resources**
There have been some small scale observational studies of CRS and behaviour:


Canadian survey (Munroe, 1999): self stimulation, continuous random purposeless movements, aggressive behaviours, lethargy, disturbances in sleep regulation, tantrums and outbursts, changes in attention span.

Danish survey (Laustrup & Gosmer, 2003): attention problems, memory impairments, self stimulation, self injurious behaviours, aggressive behaviors, reduced tolerance to changes in routines, sleeping disorders, emotional lability, lowered frustration threshold, loss of energy.

**8 CRS and behaviour**
Chris Fuggle, Sense East and Ian Noble, Sense Scotland for the CRS Steering Group

**Summary 8**
Challenging behaviour is not what defines someone – we all behave in challenging ways from time to time – we lose our tempers, or sulk, and we would hate to be labelled as ‘that person who is always shouting’ or ‘the grumpy one’. Our behaviour at those times is only an expression of the way we felt at that moment, and a way of communicating our feelings or needs there and then. People with CRS are no different – the only difference is that we label some of their behaviour as challenging because it carries risk and because we find it hard to cope with.
There are many possible causes of challenging behaviour – a particular incident will be caused by many factors so we cannot explain it as resulting from one factor or trigger. Though it can be a long and complex task to unravel why someone currently uses challenging behaviours, it is well worth the effort as by doing this we can build effective support.

9 Case study: Lesley
Heather McCrae, Sense Scotland

Heather told us about Lesley with whom she has worked for ten years – and whom she has known for 15 years. Lesley has CRS and has just turned 40. Through her life, Lesley has lived in different environments and experienced a range of approaches.

As a child, Lesley didn’t fit into either a school for deaf children or a school for blind children so she was sent to a special school, from which she was expelled for challenging behaviour. As a result she was placed in a long stay hospital where she received one hour of educational input a week.

Lesley did get out of the hospital, and moved to Overbridge. Here she had access to a continuing education centre where she learnt to sign and some life skills. The staff’s approach was that students should sit still and learn, which wasn’t right for Lesley. The records show a very controlling approach here – for example, they ‘gave permission’ for people to watch television, and they were “sent to their rooms if they were naughty”.

At Melville Street, where she moved to next, Lesley worked largely with the same staff, but one of them realised that they were using the wrong approach. She started using John O’Brien’s five points and together they went on a journey. Lesley was still living in the wrong environment for her, though, sharing with people who were messy whereas she is extremely tidy. She had been looking in estate agents' windows for years, so ...

... now Lesley has her own flat in Strathburgo, with her own front door, and is supported at last to live her own life. There has been a complete shift in the balance of power – Lesley is much more in control, and is happy and content. She is also much more expressive, with a great sense of humour which she shares with Heather.

The staff team which works with Lesley has made a huge difference:
they changed their attitudes to let Lesley be herself and show her personality
they were willing to be led by Lesley – for example one staff member went on a bus trip with Lesley to a distant swimming pool, and when they got there Lesley refused to get off the bus so they went all the way to Falkirk and ended up looking in the windows of a school – she laughed, and it was all her idea
- they promoted Lesley’s choices
- they learnt together
- they invested their own emotions
- they didn’t overprotect.

There are still challenges:
• some staff still struggle to balance their duty of care with Lesley’s choices – for example they worry about her love of cake and her weight

• they always have to ensure that the balance of power is in Lesley’s favour – though this is less of an issue in Lesley’s case than in others’

• they have to avoid making comparisons with other service users as this leads to loss of individuality – for example, it’s not fair to negatively compare someone who prefers to sit chatting with someone who likes to be out and about in the fresh air – both are valid choices

• there’s a tendency sometimes to pathologise Lesley’s behaviour rather than to see it as part of the way Lesley is.

The staff have enabled Lesley to cope with life and to achieve what she wants to do – it’s a real success story.

10 Case study: the behaviour of Mr C
Clare Smith, Sense

Summary 10
Mr C is a middle-aged man with CRS whose behaviour began to change when he reached his early 30s and he started to self injure seriously.

Staff called in Sense West’s specialist team to try to find out why Mr C’s behaviour had changed. They carried out extensive observations and investigations, and established that there were several causes of Mr C’s behaviour, including his inability to express his needs and to respond to demands. They suspected that perhaps Mr C was learning that if he head banged, staff would respond by attending to his needs, so they looked for a better way for Mr C to express the fact that he wanted something.
They fitted a buzzer in Mr C’s room which he could operate any time he needed staff support – and when staff consistently responded immediately to the buzzer, Mr C’s use of head banging dramatically declined.

**12 Where next for Sense and rubella?**
Every delegate was asked to fill in a feedback form so that we could gauge the issues which people felt were most important after two intensive days of discussion and learning. Some themes cropped up over and over – the prime one being the importance of communication between people with CRS, colleagues, families and professionals and between international practitioners and researchers.

The majority of the delegates were practitioners, and easily their main interest was to ensure that they knew about the medical effects of CRS and their implications for people with CRS.

They also noted the importance of making observations before intervening, and the usefulness of videos when making those observations. They commented on the importance of observing and reflecting before going to the doctors with a problem so as to have all the facts at hand.

Like everyone else, the small group of delegates who had CRS commented on the importance of collaboration and sharing between professions. They also noted that people with CRS have human rights, and that we need to remember that not all people with CRS have additional learning disabilities or profound sensory difficulties.

Delegates were asked what Sense should do next. The largest number felt that priority should be given to disseminating information to staff on
the ground. Many felt that Sense should also disseminate information to healthcare professionals and build working relationships with them. We need to use clinical information when we work with these health professionals – working in a Sense house can be very repetitive and staff can sometimes lose sight of their professional role so we need to enable staff to use clinical information and to be professional. Some delegates suggested that staff should be trained as experts in deafblindness as a specialist discipline.

Many people suggested ways of sharing information – for example by keeping our knowledge and websites up to date and by taking responsibility to remember to communicate our expertise in working with deafblind people to medical practitioners, so that staff are taken seriously and are seen as one important part of the individual’s health promotion team.

There was a widespread feeling that more research is needed and that we need to get better at writing up what we do in the UK. Maybe we need to allocate time within people’s jobs for doing just this? We need to find out whether the symptoms and behaviours we see are a result of CRS or just part of normal life as people get older. And we need to give priority to helping people to live healthily, enabling them to enjoy rich and fulfilling lives.

Bob Snow rounded off this summing up by saying that everyone clearly wants to work together and to share information. Sense is supposed to be an organisation for deafblind people so we’d better get moving – it took three and a half years to organise this seminar, so we need to get faster!

‘It’s not new interventions that change behaviour – it’s staff’s increased knowledge and understanding’
13 What work is going on in other organisations?

In this session, delegates from outside Sense briefly summarised the work they were doing.

DbI and the rubella network (William Green and Linda Long)
DbI is rich in human resources – it’s a voluntary organisation established 40 years ago. It’s a networking organisation bringing together people in 80 countries and fertilising cross-border co-operation. There are 12 networks within DbI such as a sibling network, a communication network and the rubella network.

Sense is an active member of DbI.

Much of the work in DbI is done in people’s free time though there is funding available for research into rare illnesses so people shouldn’t be doing it at weekends.

DbI would like the rubella network to identify areas of research which will help our health detectives on the ground.

There are European and world conferences coming up – details are on the DbI website.

It’s worth looking at the link to the Helen Keller institute. Nancy O’Donnell is there, who moderates the rubella network. There are in fact two networks – one linking people with CRS all over the world, and the other bringing professionals together.
Sense Scotland (Paul Hart)
Dr Joe Gibson did his PhD on communication using the outdoors. Now there’s new lottery-funded research. Dundee and Bergen Universities are looking at the impact of infant development in CRS.

Scotland has the whole range of services and ancillary services – they do a lot of work outdoors, visual arts, sculpting, woodwork, etc. It’s easy to get caught up in fancy activities – but it’s the ordinary ones that make the difference.

Anne Sullivan Centre, Dublin (Finola Loughney)
This is a relatively small service set up in 1995 by parents. They have a built resource for a small group of deafblind adults living in three houses. They live more in the community now than they used to.

They feel that they have lots of experience but not a huge amount of expertise.

They are developing a service for children: in Ireland children will end up in a general service if nothing is provided.

There’s not much by way of multi-disciplinary work yet but they are building up links with local contacts and have good working relationships.
Canadian Deafblind and Rubella Organisation (Denise Drake and Diane Wilcock)

They have no immediate plans to follow up their 1990s project though they would be open to partnerships and international collaborations.

Their current project is to build a database to help monitor the health and wellbeing of the people they support.

In 2011 they hope to develop a project on the lines of Sense Scotland’s Touchbase project.

Viataal, Netherlands (Trees van Nunen and Anneka Schoenmaker)

Since 2005 Trees and Anneka have been working on a long-term investigation into the effects of CRS in older people based on firm data rather than anecdotal evidence. They are looking at 70 residents with CRS who live with Viataal and with Bartimeus (an organisation for blind people).

The research has two prongs: medical and psychological. The medical assessments are relatively straightforward and are rechecked every three years. They have only just begun the psychological assessments, which are more complex.

They have created a file for each year of the study to enable them to compare data over a long period. The records note quantitative data and show degrees of increase in behaviours. They look for the effects of changes such as moving home or changing staff. There are many variables.

Eventually they want to combine the medical and psychological data.
The big difference between this work and that done in the Canadian study is that they are seeing the people in person – the data isn’t derived from checklists.

They are keen to form a research group and are more than willing to share their inventory so that others can record data too.

**Sense (Joff McGill)**
Sense is undertaking some research with the people with CRS who we directly support. We have health monitoring in place this research is an opportunity to collate and analyse data centrally; it may also produce an opportunity to identify funding for a bigger piece of work with external partners at a later stage.

Sense needs to continue to work with the National Congenital Rubella Surveillance Programme and inform them of any young people with CRS that we know about.

Sense has recognised that our focus on person centred work has helped us deliver the aspirations of those we support, but more needs to be done in the areas of meaningful occupation, friendships and quality of life.

The membership team supports a rubella network – a mailing list of those interested in rubella with bulletins sent out once or twice a year.

Sense is engaged in international collaboration, working with Deafblind International and other partners.
Sense fully supports the current MMR immunisation programme in the UK and works to raise awareness of the dangers of rubella and how to protect against it. Sense does not support single vaccines. We must not get complacent about rubella – CRS is a preventable disease, but only if vaccination rates remain high and stable.