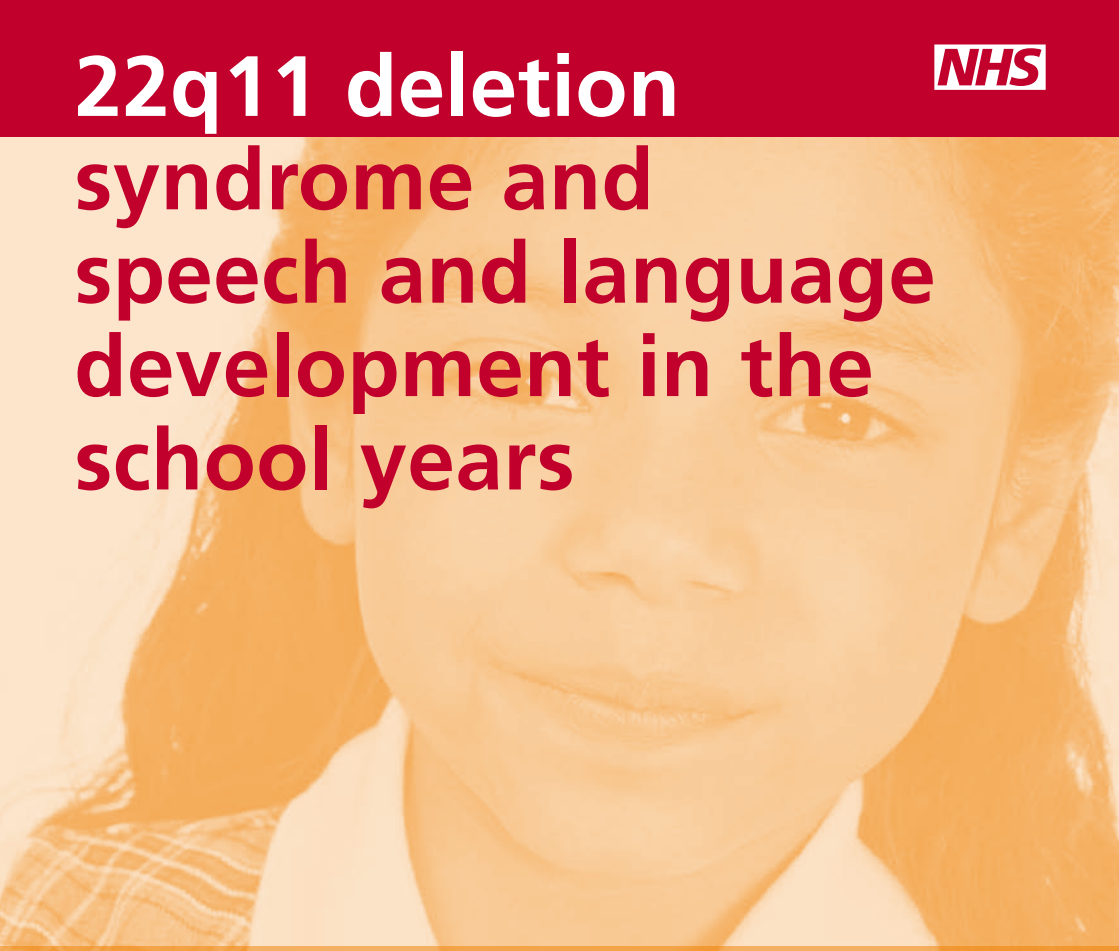
22q11 deletion syndrome and speech and language development in the school years



Information for professionals

North Thames Regional Cleft Lip and Palate Service
Great Ormond Street Hospital / St Andrew's Centre

This information sheet explains about speech and language development in children with 22q11 deletion syndrome, focussing on the school years. It gives some guidance on the most common language, communication, speech and feeding/swallowing problems that are associated with 22q11 deletion syndrome, with particular reference to how they may affect classroom, academic and social development.

What is 22q11 deletion?

22q11 deletion syndrome is a type of chromosome disorder that is found in many seemingly unrelated conditions or syndromes (collection of symptoms often seen together). Human beings have about 30 to 40,000 different genes, each of which has a function in making an individual person. The genes are arranged in pairs (one of the pair from each parent) on 23 chromosomes. Each chromosome has a long arm (referred to as q) and a short arm (p). In 22q11 deletion, a tiny part of the long arm of one of the two copies of chromosome 22 is missing at position 11.

It is diagnosed by a special blood test. You might hear the condition being called by other names as well, such as 22q11.2 micro deletion syndrome, velocardiofacial syndrome, Shprintzen syndrome or di George syndrome.

Children with 22q11 deletion syndrome can be affected in a variety of ways. They may have heart problems, reduced immunity, feeding difficulties, palate problems and difficulties with speech, language, hearing and general learning. It is important though to remember that every child is different. Please talk to the child's family and the healthcare professionals who are looking after him or her about specific concerns.

Speech and language development

It is very common for children with 22q11 deletion syndrome to have difficulties in the early years with speech and language development and most children are referred to speech and language therapy services. Some children then make good progress and catch up with their peers by school entry. However, it is very important for the school and the child's parents to keep a very close eye on their ongoing communication development, as certain difficulties only become apparent as children get older and demands made upon them change.

Language skills

Children with 22q11 deletion syndrome are often late talkers. In the early years it is quite common for them to have a better understanding of language than being able to express themselves. This gap usually gradually reduces as they approach school age and sometimes the language skills of children with 22q11 DS are similar to their peers at school in these early school years. However, the picture often changes as they get older (generally Key Stage 2 onwards), particularly around seven to eight years of age. Specific areas of difficulties start to emerge in their language skills, often coinciding with learning demands becoming more abstract and sophisticated.

Some of the specific difficulties, with ideas to help, are highlighted here:

- As understanding of language in children with 22q11 deletion syndrome is often impaired, it is important to check carefully that the child has understood what they are learning. This applies to both verbal and written material.
- Children with 22q11 deletion syndrome often have better rote reading skills than reading comprehension. It is, therefore, very important to check that they have understood what they have read.
- Children with 22q11 deletion syndrome can have problems with planning, thinking flexibly and understanding more abstract ideas. Because these skills are increasingly needed as children get older, such difficulties may not become apparent until the child is in the later stages of primary school or in secondary education. These difficulties may affect how the child processes classroom instructions and such work as reading comprehension and composition. They need help in making the link between concrete and abstract ideas not stated explicitly.
- In the classroom, children with 22q11 deletion syndrome may present with difficulties with executive function. This can include difficulties with monitoring, planning and self-initiating. Children with 22q11 deletion syndrome can therefore present as quite passive and may not let an adult know they have not understood something.
- Children with 22q11 deletion syndrome may find it difficult to hold several instructions in their memory. They generally find it easier if instructions are broken down and given in one or two steps at a time.
- Children with 22q11 deletion syndrome can have word finding difficulties and problems mastering new vocabulary. Lots of repetition is helpful. It is also worth remembering that some words can mean one thing in one context and another in a

different context. A child with 22q11 deletion syndrome may find it hard to appreciate this. It is important to be alert to this possibility and make sure that the child has understood the word in the context it is being used. He or she might find it helpful to make up their own dictionary where they can jot down new vocabulary and what it means. They can then refer back to this when they need to.

- Understanding abstract language and humour can also be very challenging for young people with 22q11 deletion syndrome in their everyday social interactions. It is common for these children to have difficulties in understanding idioms and sayings, such as 'it's raining cats and dogs'.
- Children with 22q11 deletion syndrome can find it difficult to master some syntactic rules, such as how we use verb tenses. This will affect both their understanding and ability to express themselves.
- Children with 22q11 deletion syndrome have a tendency to speak in quite short, simple sentences. They need help and encouragement to use longer and more complex sentences. This can apply to both their spoken and written work. Sometimes using a very structured approach in helping re-tell events can help. For example, asking them to think of 'who', 'what happened' 'when' and an 'ending' may help them structure composition tasks more easily.
- Difficulties with abstract language and humour can affect social relationships with other children and adults. Young people with 22q11 deletion syndrome may benefit from social communication group work.
- Children with 22q11 deletion syndrome often find it difficult to cope with the greater level of organisation required of them as they get older. Visual timetables can be a practical aid with everyday organisation. It may be helpful if the timetable is linked to specific activities. For example, if the child goes swimming on a Monday, as well as putting

the day on the timetable, the timetable entry can be illustrated with a picture of the swimming pool.

- It is really important that the learning of children with 22q11 deletion syndrome is supported both at school and in the home. A home-school diary can be one practical way of communicating regularly about how the child is doing.

If the child is not under the care of a speech and language therapist and you have concerns regarding his or her communication skills, a referral should be made for individual assessment and advice.

Speech skills

The development of speech sounds is often especially delayed in young children with 22q11 deletion syndrome and they have a tendency to disordered speech development (making unusual sounds). In some cases speech difficulties can persist into the school years and they require a lot of speech and language therapy intervention. If the child is showing these patterns of speech, he or she will need to be seen by a speech and language therapist. He or she may also need a referral to a regional cleft team to assess whether these difficulties are related to how the palate and back walls of the throat are working. Problems with nasal speech are common in children with 22q11 deletion syndrome. The causes of these problems with the palate closing off against the back wall of the throat may be due to many factors and so specialist assessment in a regional cleft team will be needed.

Feeding and swallowing

Preschool children with 22q11 deletion syndrome may experience feeding and swallowing problems. Although the majority of these will have resolved by the school years, a small number of children may experience behavioural feeding difficulties, problems chewing and controlling food and fluid in the mouth, and/or dysphagia. These problems may present as selective eating, poor weight gain, difficulty

chewing, coughing or choking when eating and/or drinking, and/or frequent unexplained chest infections. If you are concerned about a child's eating or drinking ability, this should be discussed with parents, so that a referral can be made for a speech and language therapy assessment. If a speech and language therapist is not currently involved with the child, referrals can be made through the child's family doctor (GP) or paediatrician.

Further support and information

It must be remembered that there is great variability in the presentation and severity of difficulties experienced by children with 22q11 deletion syndrome. It is, therefore, essential that the needs of the individual child are assessed by the appropriate professionals (for example, speech and language therapists, educational psychologists and/or clinical psychologists) in order to formulate individual support and learning targets to maximise academic potential.

For further information regarding your pupil's speech and language development please contact your local speech and language therapist or the speech and language therapist attached to your child's regional cleft team, where appropriate.

Max Appeal
Tel: 0800 389 1049 (free 24 hour answer phone service)
www.maxappeal.org.uk

VCFS Educational Foundation (US)
www.vcfsef.org

International 22q11 Deletion Syndrome Foundation (US)
www.22q.org

Notes

GOStA

Great Ormond Street and St Andrew's
North Thames Cleft Centre
North London - Essex - South and West Hertfordshire

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Compiled by the Speech and Language Therapy department at the North Thames Cleft Centre at Great Ormond Street Hospital and St Andrew's Centre, Broomfield Hospital in collaboration with the Child and Family Information Group at GOSH

Great Ormond Street Hospital for Children NHS Trust
Great Ormond Street
London WC1N 3JH

www.goshfamilies.nhs.uk www.childrenfirst.nhs.uk