

The background of the entire page is a photograph of a young girl with long, dark hair. She is looking directly at the camera with a neutral expression. She is wearing a dark-colored top and a beaded bracelet on her right wrist. The image is overlaid with a semi-transparent orange filter. The text is positioned over the top half of the image.

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

Information for families

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. The final section suggests ways that you, as a parent, can help your child's speech and language 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 to remember that every child is different. Always talk to the doctors and healthcare professionals who are looking after your child if you have any worries.

Speech and language development

A large percentage of young children with 22q11 deletion syndrome have difficulties with speech and language development. Most young children with this diagnosis are referred to a speech and language therapist for help. If your child's speech and language skills have progressed well, he or she may have been discharged by the speech and language therapists either before starting school or during the early primary school years. However, it is important that you and your child's school continue to keep a close eye on his or her language development.

Language skills

Your child may have been a late talker. In the early years, it is quite common for children with 22q11 deletion syndrome to have a better understanding of language than being able to express themselves. This gap usually gradually reduces as they approach school age. Sometimes the language skills of children with 22q11 deletion

syndrome are similar to their peers in these early school years. However, the picture can change as they get older (generally Key Stage 2 onwards), particularly from around seven to eight years of age onwards. Specific areas of difficulty can start to emerge, often coinciding with learning demands becoming more abstract and sophisticated.

Some of the possible areas of difficulty with some suggestions to help are highlighted here:

- If your child's understanding of language is behind that of their peers, he or she may experience difficulties following classroom instructions. It is important that teachers check carefully that the child has understood instructions and what they are learning.
- 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 into one or two steps at a time. Your child might find it helpful to repeat the instruction back in his or her own words

■ Children with 22q11 deletion syndrome sometimes find it hard to learn and remember new vocabulary. Providing plenty of opportunities to practise and repeat the new word is helpful. It is also worth remembering that some words can mean one thing in one context and another in a different context. It is important to make sure that your 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.

■ 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 and for teachers to be aware of possible difficulties in this area.

■ Some of the rules of grammar may be difficult for your child. He or she may need help in

understanding these, like when and how we apply past and future tenses.

■ Children with 22q11 deletion syndrome sometimes speak in quite short, simple sentences. They need help in encouraging them 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.

■ Children with 22q11 deletion syndrome can have problems with understanding more abstract ideas, such as time, money, humour. 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. He or she may need help in making the link between concrete and abstract ideas.

- Understanding abstract language can also be difficult for young people with 22q11 deletion syndrome. They benefit from adults helping them make the link between concrete and abstract language. They may need assistance in interpreting what is not stated explicitly verbally or in written work. You can extend this by pointing out such links in everyday life, such as when watching a film or reading stories. Your child might need you to explain idioms he or she comes across, such as 'it's raining cats and dogs'.
- 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 which some schools provide.
- A child with 22q11 deletion syndrome may have difficulties with activities such as monitoring work, planning, organising and self-initiation of tasks. One practical aid to organisation is

for your child to have access to a simple timetable at all times, both at home and school. Because children with 22q11 deletion syndrome can have difficulties with some time concepts, it can be helpful if the timetable is linked to specific activities. For example, your child may go swimming on a Monday, so as well as putting the day on the timetable, you could illustrate it with a picture of the swimming pool. Try to make the timetable both written and visual.

It is really important that the learning of children with 22q11 deletion syndrome is supported both at school and in the home. Try to establish good lines of communication with the school early on, alerting teachers to possible areas of difficulty. A home-school diary can be one practical way of communicating regularly about how your child is doing. If you are worried about how your child is progressing, discuss your concerns early with your child's teacher or the school's special educational needs co-ordinator. If your child is under the care of a speech and language

therapist, you can also discuss any concerns you have regarding his or her language and speech with your therapist. If your child is not under the care of a speech and language therapist and you have concerns regarding his or her communication skills, discuss referral with the school or your local health authority.

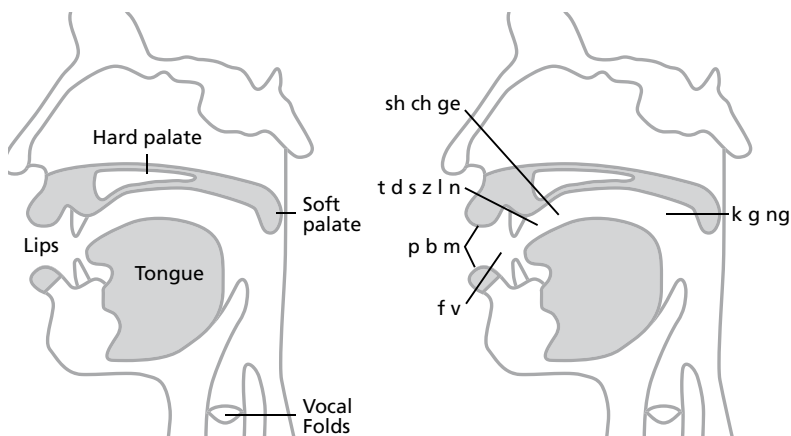
Speech skills

The development of speech sounds (how a child pronounces words) 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 in these

cases the child often requires a lot of speech and language therapy. If your 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 and side walls of the throat are working.

Nasal speech

We produce speech by air coming from the lungs, through the vocal folds (voice box) and out of the mouth. We shape the sound with our tongue, lips and teeth to say words and sounds. The diagrams below show the main parts of the speech mechanism and where consonant sounds are made.



The soft palate plays an important role in helping us talk, as it moves to close off the nose from the mouth during speech. Most sounds we use in the English language are made with the palate raised (with the nose cavity closed off) resulting in 'oral' sounds (p, b, t, d, k, g, f, v, s, z, sh, ch, ge). In English there are three sounds "m, n and ng" where the palate is lowered, so that there is a connection between the mouth and the nose resulting in 'nasal' sounds.

Problems with nasal speech are particularly common in children with 22q11 deletion syndrome. The causes of these problems with the soft palate closing off against the back and side walls of the throat may be due to many factors and so specialist assessment in a regional cleft team is needed.

When children have nasal speech, the overall sound of the voice is nasal. Nasal air emission (where air can be heard escaping through the nose during speech) or nasal turbulence (a more noisy form of nasal air emission), may also be present. It is important to find out why. Your child may need to

have two simple investigations – a videofluoroscopy (x-ray while speaking) and a nasendoscopy (a small camera passed through one side of the nose). Please see a copy of our *Investigations of nasal speech* information sheet for further information. These tests show how the soft palate and back and sides of the throat work during speech. The speech and language therapists may also make speech/audio/video recordings and sometimes measure the sound of your child's voice with a computer. These investigations will help your child's surgeon and speech and language therapists decide on the best treatment to solve this problem, which can include surgery.

Feeding and swallowing difficulties

Although feeding difficulties are relatively common during the early years, most have resolved by school age. However, in a smaller number of children, there may be persistent difficulties, most commonly limited food intake or food selection, or difficulties surrounding mealtimes, often referred to as 'behavioural

feeding difficulties'. A very small number of children may continue to experience swallowing difficulties, such as coughing or choking when eating or drinking, as well as frequent unexplained chest infections or chestiness. If you are concerned about any of these problems, contact your local speech and language therapy team, or discuss these problems with your family doctor (GP) or paediatrician.

Further information

For further information regarding your child'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



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