

Preventing infection in children with Down's syndrome

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Why should we worry about infections in children with Down's syndrome?

Children with Down's syndrome are particularly susceptible to some infections when compared to other children. Infections they are most likely to get include infections of the upper airways such as ear infections or sinusitis, of the lower airways including chest infections, bronchiolitis and pneumonia, and skin and periodontal (affecting the gums) infections.

Not only are children with Down's syndrome more likely to get these infections but when they do so there is a greater chance of them becoming seriously unwell. They are more likely than the general population to be admitted to hospital with an infection, more likely to need intensive care and to have longer hospital stays. Although the vast majority of children will respond well to treatment for their infections, the mortality rate for infection in children with Down's syndrome is higher than in the general population.

Why are infections more of a problem in children with Down's syndrome?

Children with Down's syndrome are more susceptible to infections because of a number of different factors. This includes differences in their immune system, differences in body structures and in relation to some particular illnesses that more commonly occur in people with Down's syndrome.

Immune system factors

The immune system, which is the body's mechanism for fighting infection, works less effectively

in many individuals with Down's syndrome. There may be lower levels of the white blood cells which fight infection and the body may produce lower levels of antibodies against some infections. Despite this, most of the time the child with Down's syndrome will be able to fight infections effectively, and build up immunity to infections they have been exposed to naturally and through immunisations.

Differences in body structure

Children with Down's syndrome have differences in the structure of various parts of their body. The upper airways may be quite narrow, and the mid face structures less well developed, predisposing them to upper airway infections such as ear infection, sinusitis and croup. Differences in the formation of the lower airways and lungs make respiratory infections such as bronchiolitis and pneumonia more likely.

About half of all children with Down's syndrome will have differences in the structure of the heart (congenital heart disease). Often this will affect blood flow to the lungs making respiratory infection more likely. Many of the children who have congenital heart disease will require surgery to correct the problem, in early childhood. This does however bring with it a risk of infection around the time of surgery.

Hypotonia or low muscle tone commonly occurs in children with Down's syndrome and can also make infection more likely. Difficulties with controlling the muscles of the mouth and throat may make feeding and swallowing uncoordinated, with the risk of aspiration or fluids spilling into

the airways. This makes respiratory infections more likely. Similarly gastroesophageal reflux commonly occurs in young children with Down's syndrome and regurgitated gastric fluid can also spill over into the lungs, leading to respiratory infection.

Other illnesses associated with Down's syndrome

There are a number of other medical conditions that are more likely to occur in people with Down's syndrome and can also be associated with increasing the susceptibility to infection. These include diabetes and blood disorders.

Any short-term illness or longer term health condition that can lead to poor nutrition can also make people particularly vulnerable to infection. Nutritional difficulties can be associated with many of the problems described above.

What can we do about it?

The good news is that there are a lot of things that parents, carers and health professionals can do to help prevent infection in children with Down's syndrome.

Good hygiene

Good general standards of hygiene in the home and care settings, as well as personal hygiene can help prevent skin infection in particular. Good dental hygiene including tooth brushing will help prevent gum disease.

Avoiding contact with infections

We are all unknowingly in contact with infections in our day to day lives and this helps us build up our immunity. However, it is worth avoiding contact with people who you know to be infectious.

Nutrition

A healthy balanced diet, and appropriate weight gain are important in preventing infection. This can sometimes be a challenge in children with other health problems and feeding difficulties.

Iron and vitamin D deficiency are both common in young children with Down's syndrome. Checking for this, and giving supplements where necessary is appropriate.

Immunisation

It is strongly recommended that children with Down's syndrome have all the usual childhood immunisations. Additional vaccines should also be considered. This may include:

- annual influenza vaccine (this is routinely given as nasal spray for children in the UK from the age of 2, but may also be given as an injection from 6 months of age);
- Pneumovax II protects against some types of bacterial pneumonia (the usual UK childhood vaccine schedule does include vaccines against the same group of bacteria but Pneumovax offers protection against more strains of this bacteria);

- monthly injections against RSV (respiratory syncytial virus) one of the viruses causing bronchiolitis may also be considered for those waiting for surgery for certain heart problems, or those requiring long term oxygen therapy.

Most children will make an adequate response to their immunisations, but the response rate is not 100% and it may be worth checking if they have responded through a blood test following their course of immunisations. Some vaccines can be repeated if there is evidence of an inadequate response.

Investigating and treating problems that may predispose to infection

All children get infections from time to time. However, for those who have frequent or severe infections it is worth considering whether there may be an underlying cause. Some of the tests that may be considered include:

- Blood tests to check the immune system is working properly;
- Blood tests to check nutritional status (e.g. Iron and vitamin D);
- Sleep studies to look for upper airway obstruction in sleep;

- Ph/impedance studies to check for gastroesophageal reflux;
- Urine test for diabetes.

If any condition that may make infection more likely is found it should then be promptly treated.

Prophylactic antibiotics

For children who are particularly vulnerable to respiratory infections your doctor may suggest they go on prophylactic antibiotics. This is usually at a lower dose than would be used for treating an infection, and continued long term e.g right throughout the winter months or for several years, while the child remains vulnerable

Early recognition and prompt treatment of potential infections

It is important that people caring for people with Down's syndrome, including health professionals, as well as their parents and carers are aware of the vulnerability of children with Down's syndrome to infection, recognise the signs of possible infection early, and treat promptly – see linked article below on recognising illness in children with Down's syndrome.

Recognition of Serious Illness in Children with Down's syndrome

Dr Liz Herrievan, Consultant in Paediatric Emergency Medicine (and mother to Amy, 13, who happens to have Down's syndrome)

We know that children with Down's syndrome are more likely than other children to become unwell. When they do, they tend to get more ill more quickly. We also know that the quicker a serious illness is recognised, the quicker it can be treated and the better the outcome. However, we also know that it can be really difficult, for doctors as well as families, to know whether an illness is serious or not, particularly

in children with Down's syndrome.

One of the reasons for this is that children (whether they have an extra chromosome or not) can be very good at compensating for an illness and can actually appear to be ok until their defences are overwhelmed. Children's hearts are great at adapting to an illness, beating faster to ensure an adequate circulation. That circulation is also

excellent, diverting blood to areas that need it.

Children's lungs work a little harder to ensure enough oxygen is getting in and their kidneys hold onto fluid to prevent dehydration. All this means that a child can look ok, even though an illness is taking hold. Adults are nowhere near as good at this. This compensation gives us some clues though...

faster heart rate, cool hands and feet, fewer wet nappies or faster breathing, for example.

When the body gets overwhelmed and is unable to compensate, then the child can become drowsy or lethargic, maybe very pale or ashen and may have weak breathing.

Often people worry about a temperature. A temperature can be a sign of infection, but the height of the temperature doesn't really tell us anything. Any temperature of 38°C or more in a baby under 3 months or a temperature above 39°C in a baby between 3 and 6 months is worrying, but otherwise the height of the temperature is less important than how well the child is coping with it. A child with a high temperature who is running around is better than a child without a temperature who is lethargic with cold hands and feet and difficulty breathing, for example.

Some children with Down's syndrome seem less able to control their temperature than others. These children may become cold when they're unwell, rather than hot, or their temperature may not change at all.

Some children with Down's syndrome have more trouble than others with controlling their circulation and can become very mottled or "corn beefy". Some may be prone to developing rashes at the drop of a hat and some may always seem to have a cough or snotty nose, even when they are otherwise quite well.

These things all make it more difficult for parents and health professionals to know if a child with Down's syndrome is seriously ill or not, so there is work going on to look at things known as "Soft Signs". These are things that you don't need any medical training to recognise but which might give a clue as to whether someone is ill.

Different children will have different

soft signs which are relevant to them. Things like not wanting their favourite food, not wanting to watch their favourite television programme, looking more pale than usual or being more agitated than usual may all be significant. You know your child best and this is where soft signs are important.

Another area in which families and carers can help is with supporting the doctor or nurse during any examination. The usual tricks that the doctor might use to help him or her examine a child might not work if your child has differences in understanding or sensory processing, for example, and many doctors are taught very little about learning disabilities during their training (plans are in place for this to change!). It can be really useful if you can help with distracting your child, for example, or just as helpful if you let the doctor know no amount of distraction will help! Again, you know your child best and you know what works and what doesn't for them!

There is a lot of work going on to spread awareness amongst health professionals about serious illness in children and adults with Down's syndrome and other forms of learning disability.

One area which is being highlighted is diagnostic overshadowing. This is where someone has a pre-existing diagnosis (such as Down's syndrome) and the health professional puts any new symptoms down to this diagnosis rather than looking for another cause. For example, they may assume your child is quiet and floppy because they have Down's syndrome, rather than looking for an infection.

The only ways around diagnostic overshadowing are for health professionals to realise they are at risk of it and for families and carers to point out when behaviours are normal for their child and when they are not.

Sepsis is an issue which worries a lot

of people – families and professionals alike. There is no one symptom or sign that tells us someone has sepsis and there is no one test that gives us the answer either. Sepsis is when the body over-reacts to an infection and it's very difficult to diagnose.

Sepsis can also develop during the course of an illness, so it might be that a child is seen by a doctor who diagnoses a viral infection, for example, but later the child develops sepsis. It doesn't necessarily mean the doctor missed it, it maybe it wasn't there initially. There are some "Red Flags" which can help us to diagnose sepsis, but none of them are perfect. These are:

- Not responding normally
- Not waking up properly or being unable to stay awake
- Not interested in doing anything
- Weak, high-pitched or continuous cry
- Grunting or bleating noises with every breath
- Very fast breathing or difficulty breathing
- Pauses in breathing
- Very pale, ashen or blue skin (or mottled, if this is unusual for your child)
- A rash that doesn't fade when pressed

Sometimes people are worried about calling an ambulance or may think they can get to the hospital more quickly on their own. Don't forget though, an ambulance doesn't just provide transport but can bring expert help, along with oxygen and other drugs and equipment.

Things which absolutely need an ambulance, for example, are:

- Severe difficulty breathing;
- Not breathing;
- A fit or convulsion;
- Being unconscious.

Don't forget to trust your gut instinct. This has nothing to do with your bowels, and everything to do with how well you know your child and recognising something isn't right, even if you don't know what that something is.

Down's syndrome infographic: https://docs.wixstatic.com/ugd/bbd630_66493959a5824ca5a59514207a38d8d3.pdf

Stop and Watch (NHS Cumbria) – one version of Soft Signs: <http://www.northcumbriaccg.nhs.uk/about-us/safeguarding/stop-and-watch.aspx>

Most kids with Down's Syndrome (trisomy 21) have some physiological and behavioural differences that make it difficult to assess how sick they actually are.

- Poor temp control:** may not develop a fever at all, or may be hypothermic instead
- Weak immune system:** Infections that usually cause only minor illnesses can be dangerous to kids with DS. Amy (in the photo below) spent a week ventilated in PICU when she had chicken pox!
- Mettle easily:** Kids with DS have poor control of SVR, and get mottled ("corn beef") with temperature change as well as sepsis, making assessment tricky.
- Co-morbidities common:** Remember cardiac problems, GORD, colic and autism (& don't forget the drug history)
- Leukaemia is more common** and may present atypically

Top tips for triaging & treating kids with Down's Syndrome

- Ask what's normal for this individual child:** Assessing levels of alertness, responsiveness, tone etc. can all be difficult if you don't know the individual child at baseline. Ask parents: they know their child best!
- Narrow tubes, thicker mucus:** Kids with DS get more chest & ear infections, and generally produce more snot!
- Explain and reassure:** Kids with DS often have sensory processing difficulties and can be very wary of new sensations. IPR cuffs and sala probes may be terrifying. Take time to explain and reassure.
- Optimise communication strategies:** Speech & language development lags behind understanding, so kids with DS often understand more than they can express. They're often great visual learners (but have poor short-term auditory memory and fluctuating hearing loss) so use sign, pictures and gestures. Speak slowly, clearly and maintain eye contact. Allow for sensory processing delay of several seconds: don't hurry a reply

Don't panic! Just don't forget the extra chromosome

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DSMIG was established in 1996 by 17 paediatricians. It has grown to become a network of approximately 160 healthcare professionals from the UK, Republic of Ireland and further afield. Our aim is to help promote equitable provision of medical care for all people with Down syndrome in the UK and Ireland by disseminating a wide range of information about the medical aspects of the syndrome and promoting interest in its specialist management. We are a registered charity and are the only organisation to provide this type of medical-specific service in the UK. We receive no statutory funding.



www.dsmig.org.uk

You can listen to Liz talking about her family and professional life in her episode of our Shifting Perspectives podcast. Go to www.downs-syndrome.org.uk/ShiftingPerspectivesPodcast/ to listen.



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