



**Down's Syndrome  
Association**

A Registered Charity No. 1061474

## Health Series



# Neck Instability (Craniovertebral Instability)



The  
Down Syndrome  
Medical Interest  
Group

A Down's Syndrome Association publication

## Our resources and Information Team are here to help

Please see our website for up-to-date information: [www.downs-syndrome.org.uk](http://www.downs-syndrome.org.uk)

If you would like to talk about any of the issues raised in this resource, then please get in touch with our helpline by calling 0333 1212 300 or by emailing us on [info@downs-syndrome.org.uk](mailto:info@downs-syndrome.org.uk).

**Helpline Monday - Friday 10am-4pm | Telephone: 0333 1212 300**

## The Down's Syndrome Medical Interest Group (DSMIG)

This resource has been produced in collaboration with the Down's Syndrome Medical Interest Group (DSMIG).

DSMIG was launched in 1996 and is a registered charity. It is a network of healthcare professionals – mainly doctors – from the UK and Republic of Ireland whose aim is to share and disseminate information about the medical aspects of Down syndrome and to promote interest in the specialist management of the syndrome.

**Parents and those supporting people with Down's syndrome to take part in sporting activities sometimes worry because they have heard that people with Down's syndrome can have problems with neck instability.**

**This issue has caused some confusion and controversy over the years. Although it can potentially be a serious issue, this is rare and most people will have mild symptoms before dislocation occurs.**

**This resource provides information to help parents and professionals to make informed judgments about risk and most important of all to recognize new, early warning signs that need further medical assessment. There must be a balance between encouraging people with Down's syndrome to take part in and enjoy exercise whilst identifying those few individuals who may be at increased risk for dislocation so that timely investigation and if necessary, surgical intervention can be considered.**

**'Neck instability', 'Craniovertebral instability' and 'Cervical spine instability' are umbrella terms which cover both atlanto-axial instability and atlanto-occipital instability.**

## What is Neck Instability?

In people with Down's syndrome the ligaments which stabilise the joints tend to be particularly lax and this, combined with low muscle tone, results in an unusually wide range of movement at some joints. As well as affecting the ordinary limb joints, for

instance hips and ankles, laxity can also affect the complex set of joints between the head and upper neck vertebrae. One of the functions of the vertebrae in the spine is to protect the spinal cord, a thick bundle of nerves, which runs inside the spine from the base of the brain to the pelvis. The main concern about neck instability is that this increases the risk of spinal cord damage, especially if vertebrae get misaligned.

The greatest potential for excess movement of one vertebra on its neighbour and possible misalignment is right at the top of the spinal column, at the atlanto-axial joint which lies between the top first vertebra (atlas or C1) which supports the base of the skull and the second vertebra (axis or C2), or less often at the atlanto-occipital joint between the atlas vertebra and the base of the skull (see diagram) Fig 1. There is movement at these joints whenever you nod or shake your head.

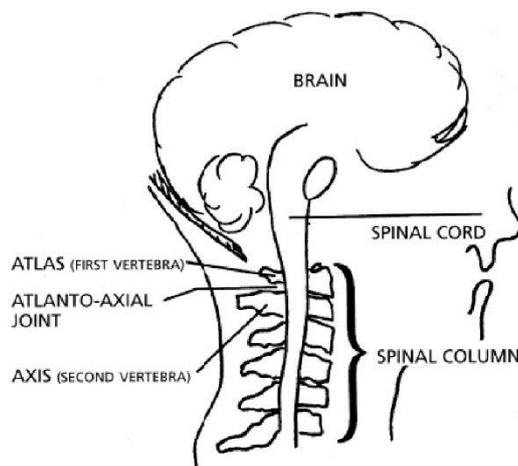


Fig 1

The axis vertebra (C2) has a central bony spur known as the odontoid peg which passes upwards from the body of the axis into the front of the spinal canal (carrying the spinal cord) in the atlas or C1. If the atlas moves too much on the axis, the odontoid peg is well placed to damage the spinal cord. (see diagram)

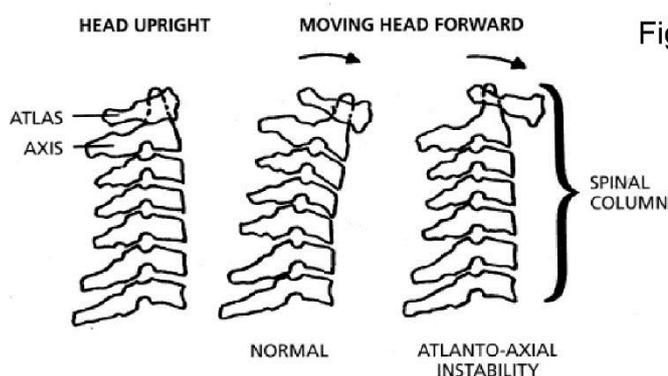


Fig 2

Fig 2 shows, in the middle picture, that when the atlas and axis are firmly bound to each other both move together when the neck bends forward. The diagram on the right shows the situation when the ligaments binding the joint are slack. Here the atlas moves forward but fails to carry the axis with it thus narrowing the spinal canal through which the spinal cord is passing. This is sometimes referred to as atlanto-axial instability or AAI. Instability and movement can also occur between the skull and first cervical vertebra so the terms neck instability, craniovertebral instability (CVI) or cervical spine instability (CSI) are now more commonly used.

## Can routine neck x-rays help predict risk in those with no symptoms?

Neck X-ray before taking part in vigorous activity has been recommended in the past (and still is in parts of the world and by Special Olympics for some sports). This advice was changed in UK in 1995 when research concluded that neck X-rays did not reliably predict risk to the spinal cord. Neck X-rays from the same person with Down's syndrome at different times can score differently, very few of the people with X-ray

evidence of neck instability ever develop any symptoms of spinal cord damage, and a normal X-ray does not mean problems due to spinal cord damage could not develop.

## What problems can be caused by neck instability?

Damage to the spinal cord in the neck can happen to anyone with or without Down's syndrome, and can cause a range of problems from mild pain or a stiff neck to paralysis in extreme cases. This can either happen suddenly as a result of a sudden shift within the joint (for example whiplash causing dislocation), or more gradually because of day-to-day pressure on the spinal cord as the neck moves. Gradual onset of symptoms due to long term instability or degenerative arthritic changes is more common in adults with Down's syndrome.

## What to look out for

Fortunately, most people have mild warning symptoms of problems in the upper spine before dislocation and long term damage occurs. **It is therefore important that ALL carers and professionals working with people with Down's syndrome are educated about warning signs of neck instability so that preventative action can be taken**

If someone you care for is showing any of these signs, they may have a problem with neck instability and should be seen urgently by a doctor:

- Pain anywhere along the neck.
- A stiff neck which doesn't get better quickly.
- Unusual head posture ("wry neck" or torticollis).
- Alteration in the way a person walks so they may appear unsteady.
- Deterioration in a person's ability to manipulate things with his/her hands.

If the onset of symptoms is sudden an emergency appointment is needed. If there is no obvious alternative explanation for these symptoms they may be related to neck instability causing nerve damage, and an X-ray in these circumstances, along with specialist referral to either an expert orthopaedic surgeon or a spinal neuro-surgeon may need to be arranged.

## What treatment is available?

If significant instability or dislocation is confirmed, and is thought to be causing problems, an operation can be done to stabilise the upper part of the spinal column and decompress any trapped nerves. The operation is delicate and is not without risk, particularly in younger children. Surgery should be performed at a specialist centre by an experienced specialist in this field. In experienced hands increasingly good outcomes are being reported. In the past there was a high rate of failure to achieve long lasting spinal fusion, however with contemporary techniques fusion is achieved in over 90% of cases.

Some children with neck instability require a period of traction to realign the joints and reduce pressure on the spinal cord before proceeding to surgical fusion. Keeping the bones immobilized after surgery whilst they heal can be difficult in children and so

surgeons may use a “halo-body jacket” to provide external support for the neck for a few weeks after surgery.

## Should people with Down’s syndrome be restricted from taking part in some sports?

The vast majority of people with Down’s syndrome do not have symptoms of neck instability and the question then is whether they should be excluded from certain activities which are associated with increased risk of neck injury?

It is clear that the very few neck injuries which have been recorded in people with Down’s syndrome whilst taking part in sporting activities were usually caused by tripping up or falling over, rather than by the sporting activity itself. In fact, the injuries recorded would have been just as likely to occur in the general population as a result of a similar fall or accident. Increased exercise and fitness may increase muscle strength in the neck and be protective. On the other hand, people with Down’s syndrome may be more at risk in some activities because they tend to be less well coordinated. These factors may well balance each other out. Therefore, there should be no justification for special anxiety when people with Down’s syndrome are taking part in everyday routine sporting activities. There is no evidence that jumping on a mini-trampoline, early stages of horse riding or simple nursery forward rolls carry more risk for a child with Down’s syndrome than any of their peers.

However higher level sports such as trampolining, diving and boxing do carry an element of risk for anyone, not just for people with Down’s syndrome, and for those with Down’s syndrome simple pre-sport screening is advisable.

The British Gymnastics Association has developed a simple screening protocol for those with Down’s-syndrome who want to take part in trampolining and other higher level gymnastics. This involves ensuring the person has good head control, full neck movement (able to “look up to the ceiling and down to floor to tie their shoes”) and has no evidence of spinal nerve damage or any of the above listed warning signs. Very few have failed this screen and been excluded from sports at this level and it is encouraging to know that of the 500 or so people with Down’s syndrome who have passed this screen no neck injuries have been sustained whilst taking part in gymnastics even at a very high level of performance. The screening protocol can be downloaded at

<https://www.british-gymnastics.org/technical-information/discipline-updates/disabilities/9316-atlanto-axial-information-pack-1/file>

## What advice can be given about other activities which have increased risk of neck injury?

It is possible that because of a tendency to instability in the neck region people with Down’s syndrome may have an increased risk of whip-lash injury following road traffic accidents. We are not sure about this, but at the moment it seems sensible to recommend that properly positioned head-rests are always in place when a person with Down’s syndrome is travelling. Similarly, after a road traffic accident it is important to

alert anyone involved at the scene to the fact that a person with Down's syndrome may be more likely to have sustained a neck injury than another person.

Another point to be aware of is that doctors will need to take special care about positioning the neck during surgery requiring a general anaesthetic. There is virtually no risk of injury if the anaesthetist and recovery room staff are alerted beforehand to the fact that the person has Down's syndrome and take the necessary care.

## References

Further references and information can be found at the website of the Down's Syndrome Medical Interest Group

<https://www.dsmig.org.uk/information-resources/by-topic/cervical-spine-disorders-craniovertebral-instability/>

## Annual health checks for people with Down's syndrome (aged 14 years plus)

In the past people with learning disabilities have not had equal access to healthcare compared to the general population. This, amongst other reasons, has given rise to poorer mental and physical health and a lower life expectancy for people with learning disabilities. Free annual health checks for adults with learning disabilities, with their GP, were introduced in 2008 as a way to improve people's quality of life.

The annual health check for people with learning disabilities is a Directed Enhanced Service (DES). This is a special service or activity provided by GP practices that has been negotiated nationally. Practices can choose whether or not to provide this service. The Learning Disability DES was introduced to improve healthcare and provide annual health checks for adults on the local authority learning disability register. To participate in this DES, staff from the GP practice need to attend a multi-professional education session run by their local Trust. The GP practice is then paid a sum of money for every annual health check undertaken.

### Who can have one?

Annual health checks have been extended to include anyone with learning disabilities aged 14 years or above. So anyone with Down's syndrome aged 14 years or over can have an annual health check.

### The benefits of annual health checks

- additional support to get the right healthcare
- increased chance of detecting unmet, unrecognised and potentially treatable health conditions
- action can be taken to address these health needs.

## How to get an annual health check

- The GP may get in touch with the person with Down's syndrome to offer an annual health check but this doesn't always happen.
- A person with Down's syndrome and/or a supporter can ask their GP for an annual health check. You do not need to be known to social services to ask for an annual health check.

Not all GPs do annual health checks for people with learning disabilities but they should be able to provide details of other GPs in your area who offer this service.

## What happens next?

- The GP practice may send out a pre-check questionnaire to be filled out before the annual health check takes place.
- The GP may arrange for the person with Down's syndrome to have a routine blood test a week or so before the annual health check.

## Who attends the annual health check?

If the person with Down's syndrome (age 16 years or over) has capacity and gives their consent, a parent or supporter can attend the health check as well.

## How long should an annual health check be?

Guidance from the Royal College of GPs suggests half an hour with your GP and half an hour with the Practice nurse.

## What areas of health should be looked at as part of the annual health check?

We have produced a check list for GPs which contains information about what should be included as part of a comprehensive and thorough annual health check. This includes a list of checks that everyone with a learning disability should undergo as part of an annual health check and a list of checks specific to people with Down's syndrome. You can find the health check list at the 'annual health checks' section of our website under 'families and carers and 'health and wellbeing'.

## What happens after the annual health check?

Your GP should tell you what they and the nurse have found during the annual health check. You should have a chance to ask any questions you have. Your GP may refer you to specialist services for further tests as appropriate. Your GP should use what they have found during your annual health check to produce a health action plan. This should set out the key actions agreed with you and (where applicable) your parent or carer during the annual health check. Your GP has to do this as part of the annual health check service.

## Information about health issues for GPs

There is information at our website for GPs about some of the more common health conditions seen in people with Down's syndrome. You will find this information at the 'annual health checks' section of our website under 'families and carers and 'health and wellbeing'.

### GPs learning disability register

People with learning disabilities experience poorer health compared to the rest of the population, but some ill health is preventable. Over one million people in the UK have a learning disability but only 200,000 are on their GPs learning disability register.

We know that people with a learning disability often have difficulties accessing health services and face inequalities in the service they receive. The Government is asking parents and supporters to speak to their GP and ensure their sons/daughters or the people whom they support are registered. It is hoped that this drive will ensure better and more person centered health care for people with learning disabilities.

The Learning Disability Register is a record of people with a learning disability who are registered with each GP practice. The Register is sometimes referred to as the Quality Outcomes Framework (QOF) Register.

If you are not sure you are on the Register, you can ask the receptionist at your GP Practice to check for you.

The doctor may have made a note on the record that a person has Down's syndrome but this does not automatically mean they have been put on the Register. When you speak to the GP about being registered, the needs and support of the person in health settings can be discussed. This information can be entered on the person's Summary Care Record (SCR) so that all health professionals at the practice know about their needs and how best to support them.

If the person is over 16 years of age or older, they must give their consent (see section in this resource about the Mental Capacity Act 2005):

- for information about their support needs to be added to their SCR
- to which information can be shared and with whom

**It's never too early (or late) to join your GP's Learning Disability Register; you can join at any age. It's a good idea for children with a learning disability to join the learning disability register at an early age. This means adjustments and support can be put in place before they reach adult services.**

## Reasonable adjustments in health care

You may have heard of the term 'reasonable adjustments' and wondered what it meant. Since the Disability Discrimination Act (1995) and the Equality Act (2010) (this does not apply to Northern Ireland) public services are required by law to make reasonable adjustments to help remove barriers faced by people with disabilities when trying to use a service. The duty under the Equality Act to make reasonable adjustments applies if you are placed at a substantial disadvantage because of your disability compared to people without a disability or who don't have the same disability as you.

So for people with physical disabilities reasonable adjustments may include changes to the environment like ramps for the ease of wheelchair users. For people with learning disabilities 'reasonable adjustments' may include easy read information, longer appointments, clearer signs at the practice, help to make decisions, changes to policies, procedures and staff training.

**If a patient with Down's syndrome is NOT on their GP's Learning Disability Register then reasonable adjustments to care for that person cannot be anticipated and made.**

Reviewed and updated 2018

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Acknowledgement to Dr Jennifer Dennis who co-authored the original resource in 2013

Publication date 2013

The Down's Syndrome Association provides information and support on all aspects of living with Down's syndrome.

We also work to champion the rights of people with Down's syndrome, by campaigning for change and challenging discrimination.

A wide range of Down's Syndrome Association publications can be downloaded free of charge from our website.

## Contact us

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