

# Neck Instability (Craniovertebral Instability)



A GUIDE FOR PARENTS AND CARERS



**A Down's Syndrome Association Publication**

# Neck Instability (Craniovertebral Instability)

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## The Health Series

The Down's Syndrome Association (DSA) Health Series covers a range of health and medical conditions written for parents and carers. The authors are professionals and specialists within each field. The DSA answers questions on a range of medical conditions and the titles in this series reflect those most commonly asked about.

**All publications in the series are available as downloads from the DSA website.**

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- 8 Oral Health Care
- 9 Continuing Pregnancy with a Diagnosis of Down's Syndrome
- 10 Depression
- 11 Thyroid Disorder
- 12 [Neck Instability](#)
- 13 Epileptic Spasms in Children
- 14 Sexual Health

## The Down's Syndrome Association (DSA)

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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## The Down Syndrome Medical Interest Group (DSMIG)

The Down Syndrome Medical Interest Group (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.

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# NECK INSTABILITY (CRANIOVERTEBRAL INSTABILITY)

**This includes both atlanto-axial and atlanto-occipital instability**

In this leaflet we will try to clarify the present understanding about risks or otherwise of neck instability (craniovertebral instability) among those with Down's syndrome.

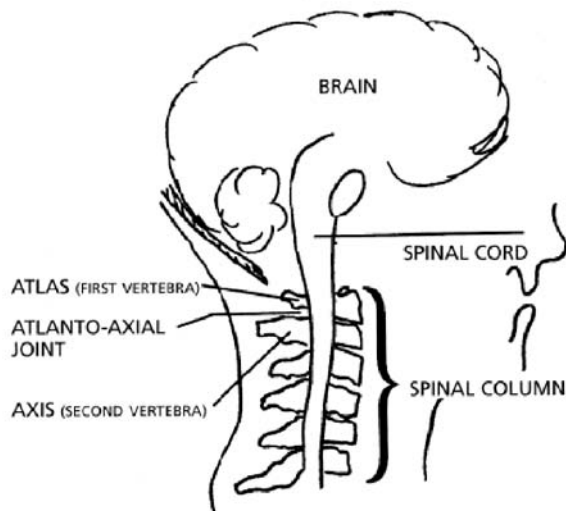


Fig 1

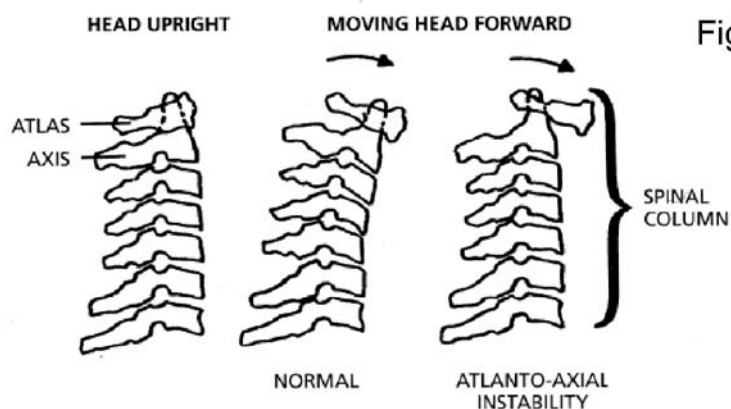


Fig 2

In the UK in 1986 a medical directive was issued mandating that all those with Down's syndrome who anticipated taking part in vigorous sporting activities must have their neck X-rayed to ensure that it was safe to do so. There were concerns that neck instability might predispose to neck dislocation and compression of the spinal cord with possible disastrous consequences.

This led to great concern about possible neck instability and its consequences for children with Down's syndrome and demands rapidly arose that all children with Down's syndrome should have neck X-rays in early childhood and possibly thereafter. There was also a tendency to limit participation in sporting and other activities.

In 1995 however the UK directive was withdrawn as it had become clear that neck X-rays did not reliably predict the risk of compression of the spinal cord and in particular that cases of spinal cord compression had very occasionally occurred in those in whom X-rays had shown no abnormality. Furthermore there was no evidence that taking part in sporting activities increased the risk of dislocation.

In the same year the American Academy of Pediatrics endorsed this view. Unfortunately however the Special Olympic Committee continues to demand neck X-rays before allowing people with Down's syndrome to compete in Special Olympic events.

## **What is Craniovertebral Instability?**

The spinal column (backbone) is a stack of specially shaped bones (vertebrae), each one moving on the one beneath. The movement occurs at a number of joints and the stability of these joints relies on the strength of the ligaments joining the bones and the tone of the muscles which straddle the bones. The spinal cord is a bundle of nerves which threads its way along a canal running through the centre of the vertebrae of the spinal column. These nerves carry information from the brain to muscles in the arms, legs and trunk (Fig 1).

In those with Down's syndrome ligaments tend to be lax and muscle tone is often low. This gives one vertebra the potential to move to a greater extent on its neighbour than is seen in the average person. As a consequence the nerve cells of the spinal cord can become distorted and injured. The greatest potential for this excess movement is right at the top of the spinal column, at the atlanto-axial joint which lies between the first vertebra (atlas) which supports the base of the skull and the second vertebra (axis), 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. The axis vertebra has a bony spur known as the odontoid peg which passes upwards from the body of the axis into the body of the atlas. If the atlas moves too much on the axis the odontoid peg is particularly well placed to cause injury to the spinal cord.

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.

The tendency for excessive movement between the atlas and axis vertebrae is known as atlanto-axial instability. If the movement is so great that one joint surface falls off the next this is known as atlanto-axial dislocation or subluxation and if this happens the risk of spinal cord damage is very high indeed. Similar problems can also occur between the base of the skull and the atlas vertebra (atlanto-occipital instability/ dislocation). Atlanto-axial and atlanto-occipital instability are known collectively as craniovertebral instability.

## **What problems can be caused by Craniovertebral Instability?**

Bruising of the spinal cord can happen to anyone with or without Down's syndrome and this can cause a range of neurological problems from mild aches and pains to paralysis.

Bruising can happen either suddenly as a result of a sudden shift within the joint, for instance following whiplash injury, or more gradually because of day-to-day abnormal pressure on the spinal cord as the neck moves. This can happen when there is chronic joint instability or arthritis or a bony abnormality.

In spite of the difference just discussed sudden neck dislocation is very rare in people with Down's syndrome but probably occurs more often than in other people. We do know however that mild symptoms occur in most people before dislocation occurs, so it is important to be on the lookout for these so that preventative action can be taken if necessary. These same symptoms may also be present in older people who don't have a major dislocation but have chronic problems due to pressure on the spinal cord. These symptoms should alert you that some compression may be occurring.

## **What should carers look out for in everyday life?**

Look out for these warning signs:

- Pain at a spot near the hard bump behind the ear.
- A stiff neck which doesn't get better quickly.
- Unusual head posture.
- 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.
- Incontinence developing in a person who has previously had no problems.

If any of these occur the person should be seen by a doctor. 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 craniovertebral instability. For the rest of this leaflet we will refer to this group as symptomatic. They may need further investigation including X-rays and specialist assessment.

## **Are routine neck X-rays helpful?**

The question which has caused the greatest confusion with regard to craniovertebral instability is whether or not people should have neck X-rays to find out if the joint is unstable. We now know that routine neck X-rays are unreliable and in those who have no symptoms no useful information will be gained by having an X-ray.

However if there are any warning signs (as above) your doctor may find DSMIG guidelines ([www.dsmig.org.uk/ publications/guidelines](http://www.dsmig.org.uk/publications/guidelines)) useful for management and further assessment. An X-ray and specialist referral may need to be arranged.

## **Are there any restrictions for people with Down's syndrome who want to take part in sporting activities?**

It is clear that very few sporting injuries have ever been recorded in people with Down's syndrome which could have been caused by craniovertebral instability. In fact the injuries recorded would have been just as likely to occur in an ordinary person as a result of a



similar fall or accident. People with or without Down's syndrome can accidentally fall onto their head whilst taking part in sporting activities. Those with Down's syndrome may be less at risk because many are less vigorous in their activities than their peers without Down's syndrome. On the other hand they may be more at risk in some activities because they tend to be less well coordinated. These two factors may well balance each other out. There is thus no justification for special anxiety when those with Down's syndrome are taking part in everyday routine sporting activities.

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 gymnastics at a higher level\* (see reference on p. 7). Very few have failed this screen and thus been excluded from sports at this level and it is encouraging to know that of the 400 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.

Doctors are often asked whether simple forward rolls or supervised bouncing up and down on a mini-trampoline in a nursery school are particularly dangerous for children with Down's syndrome. The answer is that there is no good evidence that they are. The same applies to the early stages of horse riding. At a more advanced stage in all these pursuits a greater element of risk is inevitable for everyone, whether with or without Down's syndrome.

## **Are there are other activities which may be dangerous? Can any precautions be taken?**

It is possible that because of a tendency to instability in the neck region some 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 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 when giving the sort of anaesthetic which involves passing a tube down the windpipe. 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.

## **What treatment is available?**

A doctor suspecting that craniovertebral instability may be present and causing problems in a person with Down's syndrome will probably refer that person to either an orthopaedic surgeon or a spinal neuro-surgeon. If 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. 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 and surgery can be 100% successful in treating the problem.

Children with dislocation have also been successfully treated by traction – which eases the pressure in the neck and allows the joint to get back in place – followed by immobilisation of the neck until the joint has firmed up again.

## A personal view

Our own view is that this is justifiably an area of concern for people with Down's syndrome and for their doctors, parents, carers, teachers and others who may be involved in their lives. The dilemma is, that although significant damage caused by craniovertebral instability is very rare, when it does occur it can be devastating and can result in paralysis and in extreme cases to death. Furthermore, for those who have no symptoms – which is the majority of those with Down's syndrome – there are no reliable screening tests which can identify those at increased risk.

However what we can do is be constantly aware of those warning signs which single out a person as having problems in the upper cervical spine and therefore being possibly at risk of craniovertebral dislocation. If we succeed in this people are more likely to consult a doctor before any permanent damage ensues, and the right group of people will be investigated and get early treatment. We also have to impress on doctors that this is a highly specialised area of practice and every effort should be made to ensure that those with symptoms embark on a care pathway which can lead, if necessary, to referral to a specialist national centre.

These issues have been discussed in this leaflet.

Life for everyone is not without risk. It is for the individual to decide what 'risks' are acceptable for their children or for themselves. We all have to compromise our day-to-day lives as we balance freedom to take part in and enjoy life's activities against the risk of possible injury.

## Reference

\*Atlanto-Axial Information Pack. British Gymnastics. Download from:  
[www.british-gymnastics.org](http://www.british-gymnastics.org). Available from the Down's Syndrome Association.



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