

Down's Syndrome Association Medical Series 3. ATLANTO-AXIAL INSTABILITY AMONG PEOPLE WITH DOWN'S SYNDROME Notes for parents & carers

WHAT IS ATLANTO-AXIAL INSTABILITY?

In people with Down's syndrome the ligaments which normally hold the joints stable can be very slack. This can lead to an unusually wide range of movement at some joints – much greater than in the general population. As well as affecting the ordinary limb joints this can affect the stability of one of the joints in the neck – the atlanto-axial joint. This joint is the highest joint in the spinal column and it lies just at the base of the skull. There is movement at this joint whenever you nod or shake your head (see diagrams on page 3). The lower diagram shows in the middle picture that when the atlas and axis vertebrae 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.

In some people with Down's syndrome in addition to a slack ligament the actual bones of the atlanto-axial joint may be poorly developed. Theoretically these differences could make the joint more likely to dislocate than in people without Down's syndrome.

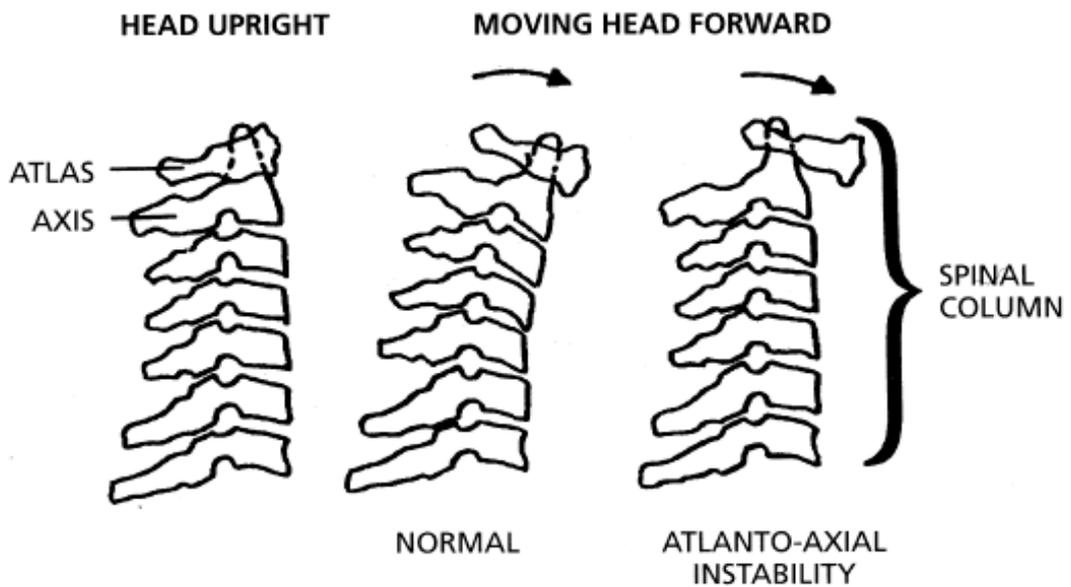
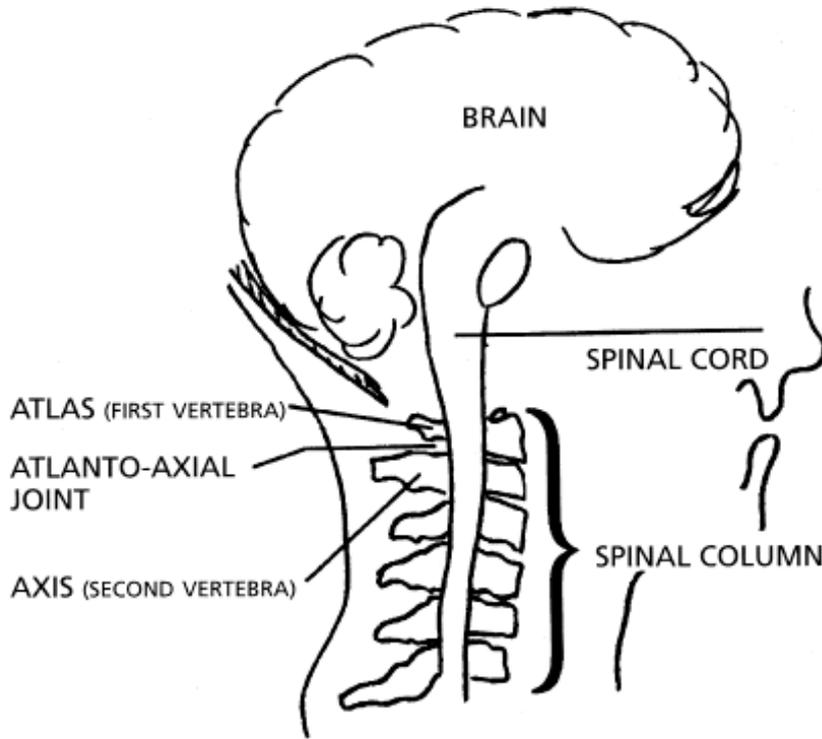
CAN ATLANTO-AXIAL INSTABILITY CAUSE PROBLEMS IN PEOPLE WITH DOWN'S SYNDROME?

The spinal cord which carries all the nerve messages from the brain to the rest of the body passes very close to the atlanto-axial joint. Bruising of the spinal cord can happen to anyone with or without Down's Syndrome. This can either happen suddenly as a result of a sudden shift within the joint or more gradually because of a day-to-day pressure on the spinal cord as the neck moves.

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Revised July 2001



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Because of the changes within the joint in some people with Down's syndrome the following questions arise:

- Are people with Down's syndrome more at risk than the rest of the population for whip-lash type injuries and possibly some sporting injuries? Such injuries could lead to paralysis.
- Are people with Down's syndrome more at risk than the rest of the population for difficulties which can arise as a result of chronic pressure on the spinal cord in the neck?
- If there is an increased risk of either of the above can we predict which people with Down's syndrome are most likely to be affected?
- Can we do anything to prevent sudden injuries or the more slowly developing long-term effects of atlanto-axial instability?

We are not sure of all the answers to these questions, which is why doctors may vary in the advice they give.

WHAT SPORTING ACTIVITIES CAN PEOPLE WITH DOWN'S SYNDROME TAKE PART IN?

It is clear that very few sporting injuries have ever been recorded in people with Down's syndrome which could have been caused by atlanto-axial 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.

Sports such as trampolining, diving and boxing do carry an element of risk for anyone, not just for people with Down's syndrome. People can accidentally fall onto their head in many sports.

People 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 may well balance each other out.

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 should be so. 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 helpers 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 when giving the sort of anaesthetic which involves passing a tube down the windpipe because in achieving this the neck position can be strained and joint dislocation can occur. There is virtually no risk of this if the anaesthetist and recovery room staff are alerted beforehand to the fact that the person has Down's syndrome.

**CAN WE PREDICT WHICH PEOPLE ARE LIKELY TO HAVE PROBLEMS
DUE TO ATLANTO-AXIAL INSTABILITY?**

What should carers look out for in everyday life?

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 look out for these.

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.

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Look out for:

- Pain at a spot near the hard bump behind the ear.
- A stiff neck which doesn't get better quickly.
- Alteration in the way a person walks so that he/she appears less good on his/her feet.
- 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.

Are routine neck x-rays helpful?

The question which has caused the greatest confusion with regard to atlanto-axial instability is whether or not people should have neck x-rays to find out if the joint is unstable.

We now know that neck x-rays taken in the routine manner followed in most x-ray departments are unlikely to be reliable in identifying instability. Some people who initially appear to have instability as shown by a neck x-ray can show no evidence of instability if re x-rayed one week later. Equally people whose neck x-ray appears entirely normal can show apparent evidence of x-ray instability one week later. On the basis of these findings there appears to be no useful information gained by carrying out such an x-ray.

X-rays can be taken in research conditions where results are known to be consistent from week to week. Even then, if instability is shown, we do not know if it is these people who are most at risk of sustaining a sudden dislocation-type injury at some time in the future. In fact we have evidence of the reverse, namely of people with no x-ray evidence of instability who subsequently sustain a neck dislocation.

What advice should be followed about x-rays?

In 1986 the Chief Medical Officer (CMO) of the then DHSS issued guidance, in the form of a letter to all doctors – CMO (86)9 – about atlanto-axial instability in people with Down's syndrome. One of the recommendations was that where vigorous sporting activity was envisaged the individual should first have their neck x-rayed in order to establish whether any instability was present. Since that time evidence has accumulated which indicates that neck x-rays do not reliably predict those at risk of compression of the spinal cord. In particular, cases

of spinal cord compression have occurred in people with apparently normal spinal cord x-rays.

The Standing Medical Advisory Committee to the CMO has now concluded that radiological examination is not a useful predictor of the risk of atlanto-axial dislocation and accordingly the CMO wrote to all doctors on 28 October 1995 withdrawing CMO (86)9. The American Academy of Paediatrics has also concluded that x-rays are not useful predictors of risk. 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 TREATMENT IS AVAILABLE?

A doctor suspecting that atlanto-axial 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 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 not without risk, particularly in younger children, but it 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

My own view is that there are two issues at the present time which are more important than whether or not we should be taking x-rays. We have to try to increase awareness among parents, carers and professionals of some possible risk situations where non-restrictive precautions can be taken to reduce the chance of neck injury. We also need to increase awareness of the symptoms of atlanto-axial dislocation and of the early signs of chronic pressure on the spinal cord. If we succeed in this, people are more likely to consult a doctor before any permanent damage ensues. Both of 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.

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Down's Syndrome Medical Interest Group Revised July 2001