

NEWS FROM THE DOWNS SYNDROME MEDICAL INTEREST GROUP (DSMIG, UK & Ireland)



A revision of the cervical spine instability (CSI) guideline

DSMIG has just revised its 1996 evidence based guideline (www.dsmig.org.uk/library/articles/guideline-cervsp-4.pdf) about issues surrounding cervical spine disorders in people with Down syndrome and I hope that this preliminary note will give some guidance to those of you who have been asking for an update. We hope to publish the revised guideline together with fully referenced background notes in the next 6 months.

In producing the revision we were very fortunate this time round to have on the steering group both Dominic Thomson, consultant paediatric spinal neurosurgeon at Great Ormond Street and Amy Walmsley from the British Gymnastics Association.

Those of you who are observant will have noticed that I am already referring to **cervical spine disorders** rather than **cervical spine instability** and that is the title of the new guideline. Although the prime focus, as before, is the diagnosis and treatment of the rare but potentially catastrophic consequences of atlanto axial instability we have widened the scope of the new guideline to draw attention to the wide range of acute and chronic neurological problems which may be caused by cervical spine disorders in Down syndrome. These may be present at any age. In childhood the incidence is low and the predominant issue is that of craniovertebral instability and here again you will notice a change of terminology. It is important to consider not only atlantoaxial but also occipito-atlanto subluxation. The risk of other problems increases with age as spinal cord compression due to premature degenerative changes becomes an additional underlying mechanism.

Much of the substance of the guideline is as before. In particular:

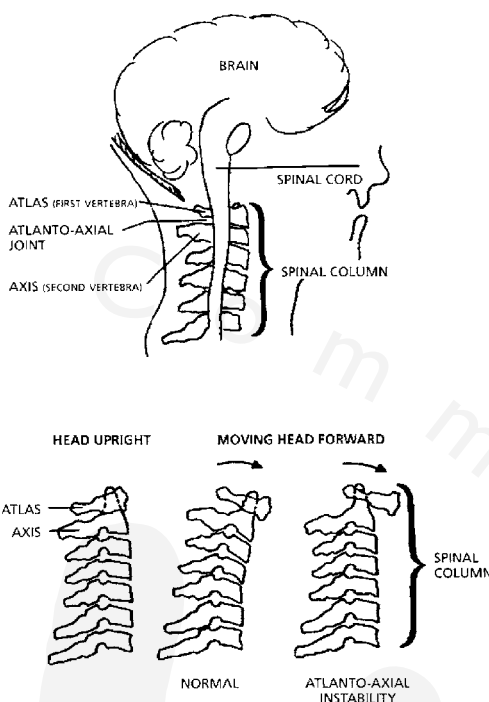
- There is no evidence of benefit afforded by radiological screening of the cervical spine in asymptomatic people with the syndrome.

One shift of emphasis has been to recognize different priorities and courses of action for those who are symptomatic and those who are not. We make the following recommendations only for those who are symptomatic.

- 'It is imperative that any person with Down syndrome presenting with new symptoms or signs that may be indicative of craniovertebral instability or myelopathy be examined and investigated expediently'
- 'In the presence of warning signs (neck pain; abnormal head posture; torticollis; reduced neck movements; deterioration of gait or frequent falls; increasing fatigability on walking; deterioration of manipulative skills) the following are required:
 - full physical and neurological examination
 - good quality flexion and extension cervical spine X Rays'

- 'Following this anyone with suspected cervical spine disorder, with or without an abnormal X Ray, should be referred to a specialist centre'

To clarify this final point. What we are saying is that if, despite possible warning signs there are no abnormal physical, neurological or radiological findings there is no need to proceed further. However if there is any one of the following - physical, neurological or radiological abnormality - referral should be made in a timely way to a specialist centre.



As to what should be included in the physical and neurological examination, that has to be left to the clinicians involved. Non-specialists may find useful the screening protocol used by the British Gymnastics Association to determine those children possibly at risk whilst taking part in gymnastics or trampolining (www.british-gymnastics.org – atlanto axial information pack). The BGA has screened over 1000 children with DS using this protocol. Only a few were considered possibly unsafe and were not allowed to proceed. The majority was cleared and many of these have taken part in gymnastics and have trampolined both recreationally and at a competitive level and there have been no untoward events. Outcome among those excluded is not known.

Following on from this we state that asymptomatic children should not be barred from sporting activities because there is no evidence that participation in sports increases the risk of cervical spine injury any more than for the general population. For specialized sports, such as trampolining, the requirements of National Governing Bodies should be observed.

Finally and importantly we stress that surgical techniques have changed since our first guideline was published in 1996. With current surgical techniques increasingly good outcomes are being reported where timely intervention is performed in experienced centres. These last 7 words are critical. There must be a high level of suspicion for risk factors and urgent referral to a specialist centre. Spinal surgeons recognize that there are steep learning curves when dealing with this particularly difficult client group and it is prudent to ensure that surgery is undertaken by those with prior experience in this area.

Dr Jennifer Dennis, Director of Information, DSMIG.

More information about DSMIG together with a wide range of medical information, including essential surveillance guidelines is available on our website –

www.dsmig.org.uk