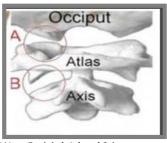
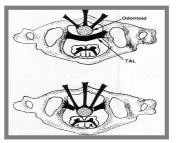
BASIC MEDICAL SURVEILLANCE ESSENTIALS FOR PEOPLE WITH DOWN SYNDROME.

Cervical Spine Disorders: Craniovertebral Instability

(Revised and Updated December 2015)

1. People with Down syndrome have a small risk for acute or chronic neurological problems caused by cervical spine/craniovertebral instability. 1-7





(A) Occipital-Atlantal Joint(B) Atlantoaxial Joint

Odontoid process stable / unstable

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- 2. Currently there are no screening procedures which can predict those at risk. In particular cervical spine x-rays in children have no predictive validity for subsequent acute dislocation/subluxation at the atlantoaxial³⁻⁷ or occipito-atlanto joint.⁸⁻¹⁴
- 3. Children with Down syndrome should not be excluded 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.^{7,12-17} In addition, children with Down syndrome should not be automatically excluded from participating in specialised sports (e.g. gymnastics). The requirements and clinical screening protocols of the relevant national governing bodies should be applied (www.british-gymnastics.org-Atlanto-Axial Information Pack).¹⁸
- 4. Prior to general anaesthesia a careful history and examination should be undertaken looking for the Red Flag warning signs below. Routine pre-operative radiography is not recommended if there are no clinical concerns. ¹⁹⁻²¹ If any child or adult with Down syndrome needs a general anaesthetic, the anaesthetist and recovery room staff must always be reminded of the diagnosis, so that appropriate care can be taken to avoid cervical injury, whilst manipulating the head and neck in the unconscious subject, although the risk of injury is small.^{4,9}
- 5. Road Traffic Accident: If a person with Down syndrome is involved in a road traffic accident personnel involved in their care should be alerted to the possibility of craniovertebral instability and of the need for particular care relative to this.^{2, 8} If there are any Red Flag warning signs detected, the patient should be immediately referred for expert opinion.
- 6. If a person with Down syndrome develops any of the Red Flags/Warning Signs below: pain behind the ear or elsewhere in the neck, abnormal head posture, torticollis, deterioration of gait, manipulative skills, or bowel and /or bladder control they should be referred immediately to an appropriate specialist ^{4,9,22} (usually a neurologist or a spinal orthopaedic surgeon) to avoid late diagnosis of Cervical Spine Instability (CSI) with potentially devastating consequences.

Red Flags/Warning Signs:

- Neck pain, or pain behind the ear
- Abnormal head posture
- Torticollis (Wry neck)
- Reduced active neck movements
- Deterioration of gait and/or frequent falls
- Increasing fatigability on walking
- Deterioration of manipulative skills
- Loss of bowel and/or bladder function

If a person with Down Syndrome develops any of these warning signs or symptoms, *passive* movements of the neck should not be undertaken.

- 7. If a good alternative explanation for the patient's symptoms is considered likely then a careful and complete clinical history, physical and neurological examination should be undertaken provided the patient has full active neck movements so that they can look up at the ceiling and down at the floor, then good quality flexion and extension cervical spine x-rays should be taken. If either clinical or radiological abnormality is found they should be referred immediately to an appropriate specialist.
- 8. It is essential that parents, relatives, carers and all healthcare professionals are made aware of these clinical/ Red Flags/Warning Signs and symptoms.

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