

screen only for chronic AAI. Nevertheless, one could argue that symptoms when they occur can be serious, such as cord compression and paraplegia in individuals who may be poor at communicating what is wrong with them. There is evidence that the earlier surgery is performed when symptoms develop, the better the results.^{17 18} The policy in our own health authority is to screen all children with Down's syndrome with a radiograph in the neutral and flexed position at the age of 4–5 years, before going into school, and maintaining particular vigilance for those found to have chronic AAI who receive repeat radiography at three to five yearly intervals in the flexed position only. Screening by radiography is relatively cheap and draws the attention of carers to neck problems in Down's syndrome, including the increased risk of acute neck injury. This radiological programme is backed up by regular checks for signs and symptoms of neck problems at a yearly medical check up.

What intervention should be provided for those found to have chronic AAI on radiography? The likely mechanism by which children with chronic AAI could increase the gap to the point when they may develop symptoms would be minor recurrent trauma causing hyperflexion of the neck. Over the course of our study, we restricted the sporting activities of those seven individuals who initially had chronic AAI, advising them against diving, trampolining, vigorous gymnastics, judo, and violent contact sports such as rugby, while encouraging such physical activities as running, jumping, swimming, and horseriding at slow speeds. None of these children deteriorated and developed symptoms. However, Cremers *et al* found that restricting sporting activities in 44 children with AAI over one year made no difference to the radiographs or clinical signs, compared with 47 with AAI who were not restricted.¹⁹ Similarly, Ohsawa *et al* reported on 14 patients with chronic AAI who remained symptom free over five years, despite lack of restriction on their sporting activities.¹¹ The evidence, therefore, suggests restriction of activity is unnecessary, especially given the rarity of symptomatic AAI. For similar reasons, radiography cannot be justified before certain sporting activities that may cause excessive flexion of the neck, although carers should remain vigilant over the possible development of symptoms. This would apply particularly to those individuals found to have chronic AAI from radiological screening at a younger age.

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Commentary

Morton *et al* present data on a large series of patients and discuss an important and controversial management problem. The paper illustrates the many difficulties of using the measurement of the atlas-dens distance on the lateral cervical spine radiograph in the management of patients with Down's syndrome.

Subluxation and instability are, in my view often, wrongly, used interchangeably. The currently accepted definitions of AAI are an atlas-dens distance of >4.0 mm¹ or >4.5 mm² and, therefore, an absolute distance of <4.0 mm in children could be regarded as 'stable'. However, it is not as easy as this because stability (or instability) is a dynamic process and an atlas-dens distance ≥ 5.0 mm which remains unchanged between flexion and extension is not an unstable atlantoaxial joint whereas an atlas-dens distance which is, say, 1.0 mm in extension and 4.0 mm in flexion is unstable. Although the majority of published data indicate that the maximal gap occurs during flexion, use of this single view gives information only about subluxation – not instability.

Morton *et al* confirm the results of previous studies,^{3 4} which have shown that the atlas-dens distance decreases with increasing age but they also confirm the finding that this change is as great, or less than, the change that can be measured on radiographs taken on the same occasion.⁵ Poor ossification of the odontoid peg, a slightly rotated film, or an uncooperative patient add to the difficulties of accurately measuring the atlas-dens distance. A reduction of AAI with increasing age is, as the authors suggest, probably due to the transverse ligament becoming firmer. However, a reduction

of this distance based on a single film taken five years later (which is what has been measured in this study) could just as easily be due to increasing ossification of the bony landmarks.

It is interesting to note that in Morton *et al*'s series neither of the two patients who developed symptoms had abnormal radiographs before becoming symptomatic and none of those with abnormal radiographs became symptomatic. The conclusion must surely be that the cervical spine radiograph is not a good clinical predictor in Down's syndrome, a conclusion which is identical to that published in this journal previously.⁵ There is no convincing evidence that the currently accepted definitions of atlantoaxial 'instability' are predictive of subluxation or dislocation with the consequent risk of neurological impairment. There is, however, good evidence that chronic dislocation is preceded by several weeks of physical symptoms and signs,⁶ with or without an accompanying history of neck trauma or an upper respiratory tract infection.

It is probably wise to obtain radiographs in flexion and extension before the first occasion

of anaesthesia or before starting a 'high risk' sport but it is probably unwise to rely too heavily on the lateral cervical radiograph, particularly if taken in only one position. Radiography has a secondary role after clinical examination and, in the presence of symptoms or neurological signs, should probably be replaced by magnetic resonance imaging or computed tomography-myelography.

STEPHEN CHAPMAN
Radiology Department,
The Children's Hospital,
Ladywood Middleway,
Birmingham B16 8ET

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