Occult spinal canal stenosis due to C-1 hypoplasia in children with Down syndrome

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Object. Little has been published about subclinical spinal canal stenosis due to C-1 hypoplasia in patients with Down syndrome. In this paper the authors performed a matched comparison study with cross-sectional survey to investigate occult spinal canal stenosis due to C-1 hypoplasia in children with Down syndrome.

Methods. A total of 102 children with Down syndrome ranging in age from 10 to 15 years were matched according to age and physique with 176 normal children. In all participants, the anteroposterior (AP) diameter of C-1 and the atlas–dens interval (ADI) were measured on plain lateral x-ray images of the cervical spine. The cross-sectional area of the atlas was also measured from a cross-sectional computed tomography image of C-1.

Results. Eight children (6.7%) with Down syndrome developed atlantoaxial subluxation associated with myelopathy. The difference in the ADI between the patients and controls was not statistically significant. The average AP diameter of the atlas and the spinal canal area along the cross-section of the atlas were significantly smaller in children with Down syndrome than those in the control group.

Conclusions. Atlantoaxial instability and occult spinal canal stenosis due to C-1 hypoplasia in patients with Down syndrome may significantly increase the risk of myelopathy. (DOI: 10.3171/PED-07/12/457)

KEY WORDS • atlantoaxial subluxation • atlas–dens interval • Down syndrome • hypoplasia of atlas • myelopathy • pediatric neurosurgery

It is well known that in children with Down syndrome, atlantoaxial subluxation associated with os odontoideum may cause neurological symptoms.3,5,8,10 However, C-1 hypoplasia has not been recognized as a risk factor for occurrence of myelopathy. Subclinical spinal canal stenosis due to a hypoplastic posterior arch of the atlas has been reported in patients with Klippel–Feil syndrome.1,12 However, little has been published on the occurrence of myelopathy related to atlantoaxial subluxation in children with Down syndrome, especially in conjunction with subclinical spinal canal stenosis due to hypoplasia of the atlas. We performed a matched comparison study with cross-sectional survey to confirm the existence of spinal canal stenosis due to hypoplastic posterior arch of the atlas in children with Down syndrome.

Abbreviations used in this paper: ADI = atlas–dens interval; AP = anteroposterior; CT = computed tomography.

Clinical Material and Methods

This study was designed as a matched comparison study with a cross-sectional survey. There were 102 children (70 boys and 32 girls) with Down syndrome who ranged in age from 10 to 15 years. These patients were age matched with 176 asymptomatic children (110 boys and 66 girls). The height and weight of children in the two groups were matched (Table 1). The asymptomatic children were children of the authors and their friends. The diagnosis of Down syndrome was made based on the characteristic clinical futures and chromosome abnormality (trisomy 21). The necks of the children were positioned carefully so that accurate lateral radiographs could be obtained. Radiographs were obtained with a constant tube-to-film and spine-to-film distance of 150 cm. In all candidates, the AP diameter of the atlas and ADI were measured from plain lateral flexion and extension dynamic x-ray images of the cervical spine. The cross-sectional area of the atlas (Fig. 1) was also measured.
from a cross-sectional CT image of the atlas. The cross-sectional area was divided into small grid cells and analyzed with a computer using an integration method. All the measurements were completed at the first examination. We obtained the informed consent from the individuals and their parents prior to the examination and obtaining radiographs and CT scans. The parents were fully aware that the data from the cases and controls would be submitted for publication, and the approval was also obtained from our institutional review board.

**Statistical Analysis**

Parametric statistical analysis was performed using Student t-test with a 95% confidence interval.

**Results**

Eight children with Down syndrome (three boys and five girls, age range 10–13 years) were identified with atlantoaxial subluxation (Table 2); an incidence of 6.7% (4.3% in boys and 15.7% in girls). Two of the eight children had reducible atlantoaxial subluxation. The condition of four patients was complicated by os odontoideum. All patients with atlantoaxial subluxation exhibited spastic gait, hyperreflexia, pathological reflex, and disturbance of finger movement. We recommended surgery to the eight patients, but we could not obtain agreement. The ADI ranged from 6 to 9 mm among the patients who exhibited atlantoaxial subluxation, and all patients exhibited hypoplasia of atlas. The mean (± standard deviation) ADI was 2.5 ± 1.0 mm in all children with Down syndrome and 2.2 ± 1.0 mm in healthy children. The difference between the two groups was not statistically significant in boys or girls. The average AP diameters of the atlas were significantly smaller in patients with Down syndrome than in controls. The results were the same in boys and girls (Table 3). The cross-sectional area of the atlas was significantly smaller in children with Down syndrome than in the control group (Table 4). Figure 2 provides an example of CT images of a patient with Down syndrome and a healthy male control of the same age. The AP diameter and cross-sectional area of the atlas were smaller in the patient with Down syndrome.

**Discussion**

Atlantoaxial dislocation in patients with Down syndrome was reported by Tischler and Martel in 1965 and by Dzenitis in 1966. Since then, many articles have appeared in the literature detailing imaging-documented atlantoaxial instability in children with Down syndrome. The present study suggests that occult spinal canal steno-

![Image](image.png)
Occult spinal canal stenosis in children with Down syndrome

sis exists in patients with Down syndrome. To our knowledge, this is the first matched-comparison study that has examined occult spinal canal stenosis due to hypoplasia of the C-1 posterior arch in patients with Down syndrome. In 1992, Martich et al. reported hypoplastic posterior arch of atlas in children with Down syndrome; however, that study was not a matched comparison, and the children were younger (2–3 years old). In our study, the hypoplasia of C-1 in children with Down syndrome was statistically significant. The pathomechanism of myelopathy in patients with Down syndrome has not been clarified. However, the occult spinal canal stenosis due to C-1 hypoplasia must be a risk factor of myelopathy for patients with Down syndrome. All patients with atlantoaxial subluxation in the current study exhibited myelopathy. The ADI among the patients who exhibited myelopathy was less than 9 mm, which did not indicate severe atlantoaxial subluxation.

Conclusions

When dealing with children with Down syndrome, it must be remembered that the patients may have occult spinal canal stenosis which can cause myelopathy.

References


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