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CT Metrizamide Myelography of the Cervical Spine in Morquio Syndrome

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Since the syndrome was first described concurrently by Morquio [1] and Brailsford [2] in 1929, radiology has played a vital role in the diagnosis of the Morquio-Brailsford syndrome and in the management of complications of the disease. Symptoms of spinal cord compression are common. They are related to dysplasia of the dens with atlantoaxial subluxation and narrowing of the spinal canal [3-5].

Spine radiographs demonstrate flattening and widening of the vertebral bodies, which may be pronounced and can involve the entire spine in the most developed form of the disease [6]. From the central position of the anterior aspect of the thoracic and lumbar vertebral bodies project peculiar beaklike processes that are characteristic of the syndrome. Anterior wedging of the vertebral bodies may result in varying degrees of kyphosis [4]. Gibbus deformity at the thoracolumbar level is common, and may cause cord compression at this level [4]. In the cervical region the flattening of the vertebral bodies results in shortening of the neck [6]. The intervertebral disk spaces retain their normal height and may be slightly thickened [7].

The odontoid process shows varying degrees of dysplasia. It is usually hypoplastic and may be absent [4, 5]. Radiographs demonstrate increased mobility at the atlantoaxial junction with flexion and extension. Chronic atlantoaxial subluxation may develop and cause severe cord compression [4].

Narrowing of the spinal canal is a well recognized finding in Morquio syndrome. This narrowing usually occurs in the anteroposterior diameter secondary to shortening of the pedicles due to premature synostosis of the centers of ossification of the body with those of the lamina [6]. Narrowing of the neural arch is most severe at the level of the first cervical vertebra. With anterior dislocation of C1, in addition to narrowing of the arch of C1, the posterior arch of the atlas herniates into the foramen magnum and may give the false impression of platybasia [4, 8].

Thickening of the dura and compression of the spinal cord and subarachnoid space by deposits of mucopolysaccharide has been described in patients with Maroteaux-Lamy syndrome (MPS-VI) and a variant of Hurler-Scheie

(MPS-1 H/S) [9, 10]. To our knowledge, this is the first report of mucopolysaccharide deposit in the dura of a patient with Morquio disease.

Subjects and Methods

During a 3 year period, four patients with Morquio syndrome treated at the Hospital for Sick Children were examined by metrizamide myelography in conjunction with computed tomography (CT). A total of six procedures were performed. Three examinations were performed on one child to monitor surgical decompression and fusion procedures. Plain films of the spine and conventional metrizamide myelography using concentrations of 170-190 mg I/ml were obtained in each case prior to CT metrizamide myelography under general anesthesia. Four of the studies were performed with an Ohio-Nuclear Delta 50 scanner, and two studies with a GE 8800 scanner with retrospective image analysis (Review).

Case Reports

Case 1

A previously healthy, apparently normal 2½-year-old girl fell from her high chair and was found flaccid in all extremities. Radiographs of the spine showed characteristic beaking of the thoracolumbar spine and the diagnosis of Morquio disease was made. Atlantoaxial subluxation was demonstrated with flexion and extension views. Metrizamide myelography revealed severe narrowing of the anteroposterior diameter of the spinal canal, worse at C1 (fig. 1A). CT metrizamide myelography showed severe sagittal narrowing of C1, measuring 10.8 mm compared with the lower limit of normal value of 14 mm. CT metrizamide myelography also demonstrated that, with extension of the neck, the posterior arch of C1 would herniate into the foramen magnum (figs. 1B-1E). At surgery, the posterior arch of C1 was found within the posterior foramen magnum, and this configuration complicated the subsequent laminectomy of C1. After surgery, the patient recovered full strength in all four extremities.

At the level of the most severe cord compression, the metrizamide column in the subarachnoid space is narrowed and may even be obliterated. The increase of metrizamide density from figure 1B, the level of greatest compression, through figure 1E, the level of least compression, illustrates this phenomenon.

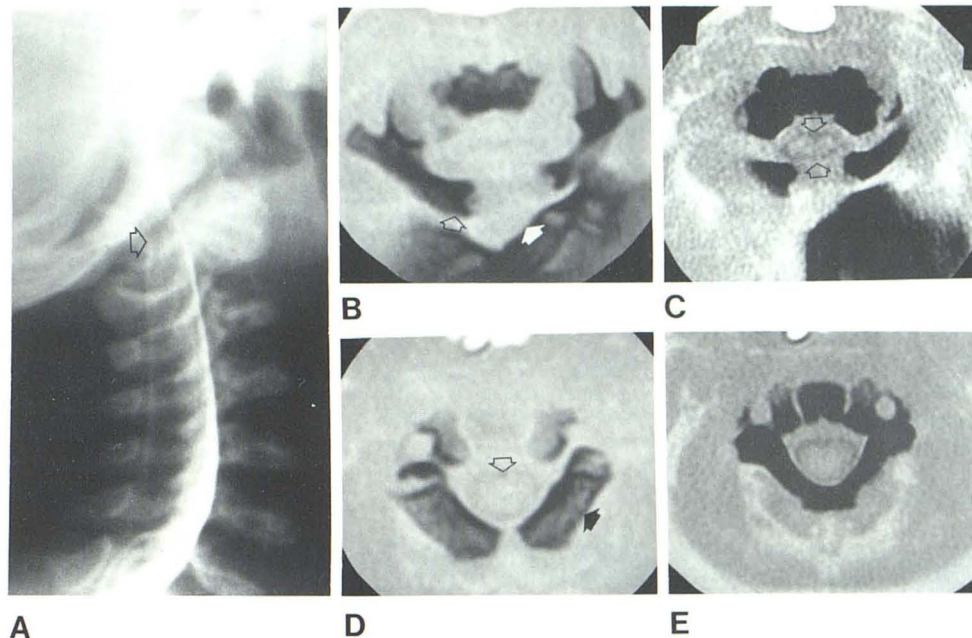
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Fig. 1.—Case 1. **A**, Lateral view of standard metrizamide myelogram. Forward subluxation of atlas causes compression of cervical cord (arrow). **B**, CT metrizamide myelogram at occipito-cervical junction. Displacement of posterior arch of C1 within foramen magnum on hyperextension (arrows). **C**, At level of atlas. Marked anteroposterior (AP) narrowing of neural arch with cord compression (arrows). **D**, At C3. Mild narrowing of spinal canal, less than at C1–C2, with thickened laminae (closed arrow) and slightly compressed subarachnoid space (open arrow). **E**, In low cervical spine. Increased density of metrizamide because of wider subarachnoid space. No cord compression.



Case 2

A 14-year-old boy with Morquio syndrome developed lower extremity weakness at age 2 years for which a T9–L5 laminectomy was performed. After surgery, he continued to have bilateral clonus and hyperactive reflexes in both lower extremities and developed enuresis and progressive quadriplegia by the age of 10 years. Spine radiographs at that time demonstrated a hypoplastic dens. CT metrizamide myelography showed severe narrowing of the cervical spinal cord by a soft-tissue mass anterior to the cord (figs. 2A and 2B). A decompressive laminectomy with posterior fossa decompression was performed. Biopsy of the mass anterior to the cord revealed mucopolysaccharide within the dura. The child recovered partial strength in his lower extremities but continued to have symptoms of a neurogenic bladder. CT metrizamide myelography was performed again and demonstrated herniation of the posterior cervical spinal cord through the laminectomy defect with lateral compression of the cord by the bony laminae. The posterior part of the C1 lamina was demonstrated within the posterior foramen magnum. The laminectomy was widened and a cervical fusion was performed from C1 to C5. The child continued to develop progressive lower extremity weakness and gait disturbance, and at age 14 years CT metrizamide myelography showed persistent herniation of the spinal cord through the laminectomy defect with mucopolysaccharide deposits anterior to the cord (fig. 2C).

Case 3

A 5-year-old boy with Morquio syndrome developed sudden right hemiparesis after a fall from his tricycle. Radiographs of the spine showed dislocation of the dens with subluxation of C1 on C2. An attempted atlantoaxial fusion was performed. CT metrizamide myelography demonstrated stenosis of the cervical spinal canal (fig. 3). There was nonunion of the attempted fusion, and a second unsuccessful fusion procedure with decompressive laminectomy was attempted the next year. Although there was continued subluxation of C1 on C2, the child's neurologic symptoms improved to the point that he was left with only mild spastic quadriparesis. At age 7 he fell

at camp and developed acute dense left hemiparesis. A decompressive laminectomy and fusion was performed from the occiput to C5. The child remained clinically unchanged except for a slight increase in lower extremity strength.

Case 4

A 17-year-old girl with Morquio disease presented at age 14 years with progressive leg weakness and decreased exercise tolerance. Radiographs of the cervical spine showed dislocation of the odontoid with atlantoaxial subluxation (figs. 4A and 4B). CT metrizamide myelography showed marked narrowing of the neural arch in the cervical region and forward displacement of the dislocated dens (figs. 4C and 4D). Fusion of the cervical spine from the occiput to C3 was performed and a part of the inferior arch of C1 was removed. After surgery the patient had an excellent recovery with no evidence of residual motor weakness and no clonus.

Discussion

It has long been recognized that patients with Morquio syndrome are predisposed to paralysis attributable to spinal cord compression. The patients may present in early childhood with difficulty walking, which usually develops into a chronic myelopathy of variable progression [11]. Many patients develop symptoms of neurogenic bladder. It is not unusual for the patients to be bedridden in their teenage years [5]. Patients with Morquio syndrome are particularly at risk for acute quadriplegia and respiratory distress after trauma, which may be relatively minor. For this reason some authors suggest prophylactic atlantoaxial fusion for patients with Morquio syndrome [5].

CT provides excellent demonstration of the dysplastic vertebral body and neural arch changes of Morquio syndrome. Severe shortening of the AP diameter of the neural

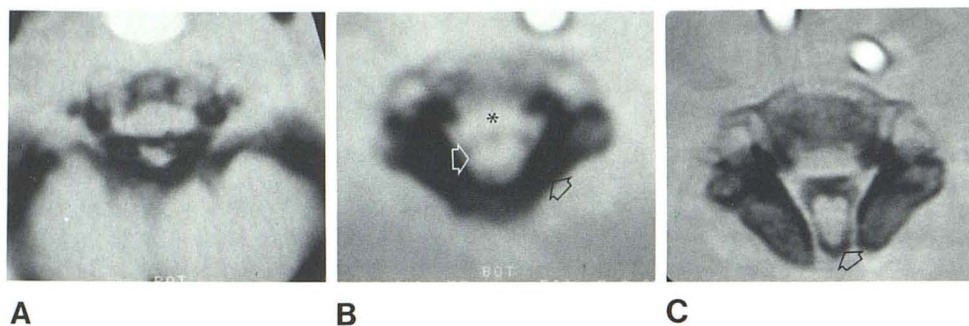


Fig. 2.—Case 2. **A**, CT metrizamide myelogram at C1. Narrowing of spinal canal most marked at C1. **B**, At C3. Thickened laminae (black arrow), compressed cervical cord (white arrow), and mucopolysaccharide deposit anterior to cord (asterisk). **C**, At C3 4 years later. Herniation of spinal cord through laminectomy defect (arrow).

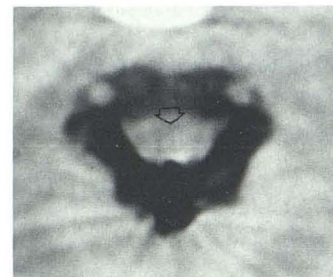


Fig. 3.—Case 3. CT metrizamide myelogram at C2. Severe cord compression with narrowed subarachnoid space (arrow). Wires from fusion procedure cast metallic artifacts.

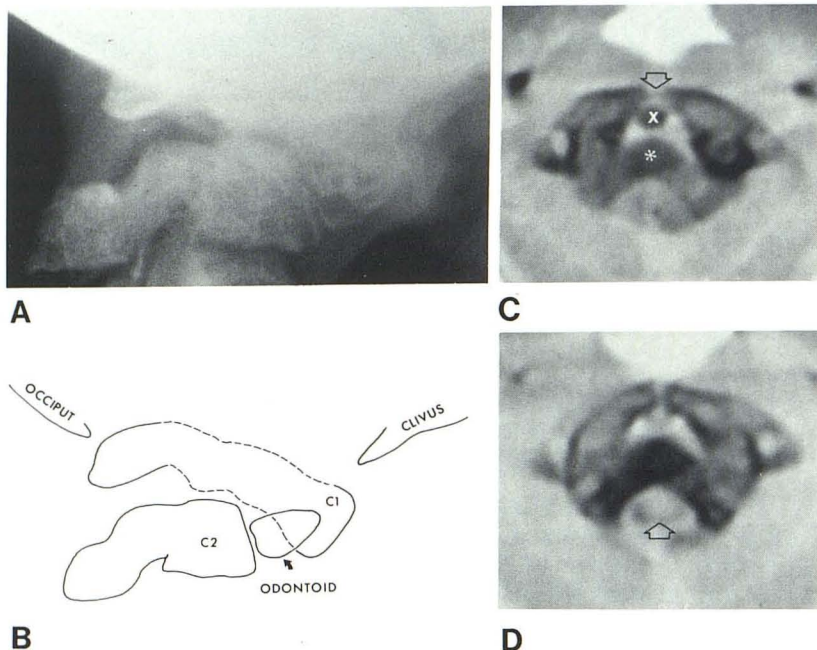


Fig. 4.—Case 4. **A** and **B**, Plain lateral cervical spine radiograph and drawing. Dislocation of dens, atlantoaxial subluxation, and upward displacement of posterior arch of C1 into foramen magnum. **C**, CT metrizamide myelogram at C1–C2. Dislocated dens (X) anterior to body of C2 (asterisk). Open anterior arch of C1 (arrow), in normal relationship with dens but showing forward subluxation with reference to body of C2. **D**, 7 mm lower. Severe compression of cord (arrow) by C1–C2 subluxation.

arch is seen with thickening of the lamina, which further compromises the area within the spinal canal. These changes were present to a variable degree among the children in our series, and appeared to be age-dependent, with the narrowing of the neural arch and thickening of the lamina most marked in the older children. These changes, quite evident on axial CT, are difficult to demonstrate with plain films.

CT metrizamide myelography provided accurate demonstration of the extent and degree of spinal cord compression in our series. In each of the four children, marked narrowing of the spinal canal was demonstrated, most severe at the level of the atlas. Spinal cord compression was caused, in part, by atlantoaxial subluxation in three cases and by deposits of mucopolysaccharide in the soft tissue anterior to the cord in one case. Two patients were shown to have

displacement of the shortened posterior arch of C1 into the posterior foramen magnum. This upward displacement of the C1 neural arch contributes to surgical difficulty in performing decompression laminectomy and cervical fusion.

The multiple cervical spine changes in Morquio syndrome may be difficult to demonstrate before surgery and even more confusing after decompressive laminectomy and fusion procedures. We have found CT metrizamide myelography extremely helpful in demonstrating the cause of spinal cord compression in Morquio disease as well as narrowing of the spinal canal, herniation of the posterior arch of C1 into the foramen magnum, and mucopolysaccharide deposits within the soft tissues anterior to the cervical spinal cord. Cord compression secondary to atlantoaxial subluxation is best demonstrated on CT metrizamide myelography, whereas the bony relationships of C1 and C2 need a lateral

projection, with sagittal reconstruction, plain films, or routine tomography. In our experience, plain CT of the spine without metrizamide does not currently provide sufficient soft-tissue resolution to accurately differentiate structures within the spinal canal.

The use of CT with metrizamide is suggested as the diagnostic procedure of choice after standard AP and lateral radiographs in this most complex disease involving the spine. CT demonstrates with ease, safety, and precision the entire spectrum of abnormalities of bone, intraspinal soft tissue, cord, and occipitocervical relationships.

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