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Opinion

Cardiac Complications of Morquio's Syndrome

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Morquio's syndrome is a rare type of mucopolysacharidosis (type iv) which becomes evident when the affected infant starts to walk. The condition is marked by severe dwarfism especially of the torso, short neck, prominent sternum, dorsolumbar kyphosis, genu valgum, flat feet and wadling gait. In contrast to hurler's syndrom, the mental retardation is slight to normal.

They get aortic valve disease, specifically aortic regurgitation.

To make the diagnosis one picks up keratan sulfate in the urine. There is a type a and a type b form of the disorder.

They also get cloudy cornea and spinal malalignment. Hypoplasia of the odontoid causes atlantoaxial subluxation, producing myelopathic changes.

In a nut-shell the internal organs continue to grow while the skeltal system stops growing. A good example of this was in the movie " the mighty freak."

Most die of congestive cardiac failure related to aortic regurgitation and compression of the heart! On auscultation they characteristically get early diastolic murmurs of aortic incompetence. All the other stigmata of a wide pulse pressure are picked up; corrigans pulse etc.

The only form of reversing the condition is by means of a cardiac transplant but the problem is that the skeletal changes are so severe.

Gene therapy may be of value as may stem cell therapy.

Most patients are put onto anti-cardiac failure therapy which includes lasix, slow k digoxin and an ace-inhibitor.

Synonyms for the condition are;

Eccentro-osteochondroplasia,