



INTERNATIONAL
MEDICAL REVIEW
ON DOWN'S SYNDROME

www.elsevier.es/sd



ORIGINAL

Advanced myelopathy in people with Down syndrome[☆]

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Received 20 July 2017; accepted 5 October 2017

KEYWORDS

Down syndrome;
Myelopathy;
Spine;
Activities of Daily
Living;
Nurick;
Incontinence

Abstract

Background: Myelopathy is a condition that significantly impacts a person's mobility and independence. In people with intellectual disabilities, such as Down syndrome, the negative impact of myelopathy is magnified. Myelopathy in Down syndrome may be related to atlanto-axial instability or degenerative pathology. Our experience with these patients has led us to hypothesize that their myelopathy is commonly undiagnosed until very severe. In this study we seek to determine whether patients with Down syndrome present with more severe myelopathy than those without Down syndrome.

Methods: We performed a retrospective medical record review of patients with Down syndrome who were treated for myelopathy by the Tufts Neurosurgical Practice. Eight patients met the criteria and were graded for severity of myelopathy on the Nurick Scale. We compared the patients with cervical spondylotic myelopathy and Down syndrome to patients who were treated for cervical spondylotic myelopathy as reported in Furlan et al. and Fehlings et al.

Results: The average Nurick grade for patients with Down syndrome was 4.2 (SD 0.84, $n=5$). The average Nurick grade as reported by Furlan et al. was 2.8 (SD 0.68, $n=81$) and by Fehlings et al. was 3.14 (SD 0.97, $n=278$). The independent samples t -test resulted in a P value < 0.000 and 0.016 with Furlan et al. and Fehlings et al. respectively.

Conclusions: The patients with Down syndrome in our study presented to neurosurgery with more severe myelopathy than patients without Down syndrome. It is important for physicians caring for people with Down syndrome to be aware of the presentation of myelopathy and consider the condition in the differential diagnosis of a Down syndrome patient with functional decline.

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[☆] A previous revision of the abstract for this research was presented to the assembled attendees at the New England Neurosurgical Society Annual Meeting on June 24, 2016.

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<https://doi.org/10.1016/j.sdeng.2017.11.001>

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PALABRAS CLAVE

Síndrome de Down;
Mielopatía;
Columna;
Actividades de la vida
diaria;
Nurick;
Incontinencia

Mielopatía avanzada en personas con síndrome de Down

Resumen

Antecedentes: La mielopatía es una enfermedad que afecta de manera significativa la movilidad y la independencia del paciente. En personas con discapacidades intelectuales como el síndrome de Down, el efecto negativo de la mielopatía se magnifica. La mielopatía en el síndrome de Down puede estar relacionada con inestabilidad atlantoaxial o enfermedad degenerativa. Nuestra experiencia con estos pacientes nos ha llevado a formular la hipótesis de que su mielopatía no se suele diagnosticar hasta que es muy grave. En este estudio nos proponemos determinar si los pacientes con síndrome de Down presentan mielopatía más grave que los que no tienen síndrome de Down.

Métodos: Llevamos a cabo una revisión retrospectiva de historiales médicos de pacientes con síndrome de Down que recibieron tratamiento para la mielopatía por parte del Tufts Neurosurgical Practice. Ocho pacientes cumplían los criterios y se les evaluó la gravedad de la mielopatía según la escala de Nurick. Comparamos a los pacientes con mielopatía cervical espondilótica con los pacientes con síndrome de Down que recibieron tratamiento para la mielopatía cervical espondilótica, según la información en Furlan et al. y Fehlings et al.

Resultados: La media del grado de Nurick para pacientes con síndrome de Down fue de 4,2 (DE 0,84, n=5). La media del grado de Nurick según la información en Furlan et al. fue de 2,8 (DE 0,68, n=81) y según Fehlings et al. fue de 3,14 (DE 0,97, n=278). Las pruebas t independientes de las muestras arrojaron un valor $p < 0,000$ y $0,016$ con Furlan et al. y Fehlings et al., respectivamente.

Conclusiones: Los pacientes con síndrome de Down de nuestro estudio acudieron a Neurocirugía con una mielopatía más grave que la de los pacientes sin síndrome de Down. Para los médicos es importante atender a personas con síndrome de Down para darse cuenta de la presentación de mielopatía y considerar la afección en el diagnóstico diferencial de un paciente con síndrome de Down y con deterioro funcional.

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Introduction

Myelopathy is a common spine condition in which the spinal cord is compressed and damaged.¹ It can be caused by acute injury, progressive degeneration, or vertebral instability. It is most often caused by disk degeneration and stenosis of the spinal canal. If untreated, narrowing of the spinal canal and compression of the cord can cause demyelination and necrosis, which are irreversible.² It can affect the cervical, thoracic, and lumbar spine. Cervical spondylotic myelopathy (CSM) predominates and worldwide is the most commonly treated pathology of the spinal cord.^{2,3} Neurosurgical intervention is indicated when myelopathy is symptomatic and progressive.¹ A favorable outcome correlates strongly with early treatment: it is important for people with myelopathy to be identified quickly because people whose disability is severe are less likely to improve from treatment.²

There are several well-studied conditions in people with Down syndrome (DS) which can cause myelopathy. One is atlanto-axial instability caused by laxity of the transverse ligament. It can lead to compression of the cervical spine.⁴ Atlanto-axial instability affects 10–20% of people with DS but is asymptomatic in 98–99% of cases.⁵ The danger posed by atlanto-axial instability to pediatric patients with DS has led to specific recommendations for cervical spine X-rays by treating physicians whenever there is a change in neurological function.⁶

CSM of the sub-axial cervical spine is common in people with DS, even if it is not commonly diagnosed. In people with DS some studies have found a 45% prevalence of moderate or severe CSM.⁷ Clumsiness and gait change are typical onset symptoms in the natural history of CSM, but symptoms of CSM can be as severe as incontinence and quadriplegia.⁸ The high prevalence of CSM in people with DS is due primarily to degenerative changes that result in stenosis of the spinal canal.⁹

Clumsiness and gait abnormality are the most common symptoms people notice at the onset of CSM.⁸ Insomuch as pain is the most motivating symptom for a person to seek treatment, myelopathy may be insidious because it is often painless. When there is pain caused by CSM, many people complain of shoulder pain or referred pain.⁸

Many people with CSM experience a decline-plateau of symptoms.¹⁰ In a typical scenario, a person might notice a new symptom, such as decreased dexterity of an extremity, which will then remain unchanged for some time. Symptoms can remain static for years; then suddenly existing symptoms become worse or a new symptom of the developing myelopathy will emerge. This step-wise feature of myelopathy can alter a person's pursuit of treatment. If an initial symptom is problematic but not severe, and has plateaued, the person may resist seeking treatment or accepting any non-conservative treatment. Upon decline in function or

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