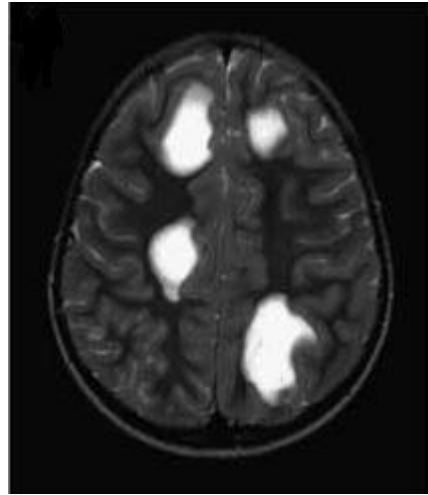


Er MS í Börnum Sami Sjúkdómur og MS í Fullorðnum

Ólafur Thorarensen Barnalæknir
Sérfræðingur í heila- og taugasjúkdómum barna
Barnaspítali Hringisns
Barn- och ungdomshabiliteringen Region Skåne





Afmýlandi bólgusjúkdómar í miðtaugakerfi barna árin 1990-2010

Brynjar Þór Guðbjörnsson



HÁSKÓLI ÍSLANDS





Original Article

Nationwide Incidence of Acquired Central Nervous System Demyelination in Icelandic Children



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ABSTRACT

INTRODUCTION: Recognizing acquired demyelinating syndromes and multiple sclerosis is important to commence early treatment. The objective of this study was to describe the incidence of acquired demyelinating syndromes and multiple sclerosis among the entire Icelandic pediatric population according to recently promoted criteria. **PATIENTS AND METHODS:** The study included all children in Iceland (<18 years) with acquired demyelinating syndromes and multiple sclerosis from 1990 to 2009 with a minimum of 5-year follow-up. Clinical data were gathered and radiological images reviewed. The cohort included all patients with acquired demyelinating syndromes and multiple sclerosis in the Icelandic pediatric population. **RESULTS:** Eighteen patients with acquired demyelinating syndromes and multiple sclerosis were included, the total annual incidence being 1.15/100,000 (acquired demyelinating syndromes 1.02 and multiple sclerosis 0.45/100,000). The median age at diagnosis was 14.25 years (range 1.25–17.5 years). Thirteen patients were initially diagnosed with clinically isolated syndrome, two had acute disseminated encephalomyelitis, two had multiple sclerosis, and one had neuromyelitis optica. Seven children were diagnosed with multiple sclerosis; three patients with clinically isolated syndrome developed multiple sclerosis after the age of 18 and were not included in the multiple sclerosis group. The gender ratio was equal. Of the nine girls, seven were diagnosed with clinically isolated syndrome. Most patients (11 of 18) were diagnosed during the period January through March. Oligoclonal bands in cerebrospinal fluid were exclusively found in patients with multiple sclerosis and clinically isolated syndrome and 13 of 14 available magnetic resonance images revealed clear abnormalities. **CONCLUSION:** The annual incidence of acquired demyelinating syndromes and multiple sclerosis in Iceland was 1.15/100,000 children. The risk of progression from clinically isolated syndrome to multiple sclerosis was high. There was no female preponderance.

Keywords: acquired demyelinating syndrome, children, multiple sclerosis, ADEM, CIS, transverse myelitis

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Introduction

Multiple sclerosis (MS) is uncommon in children and less than 10% of patients with MS are diagnosed during childhood. A diagnosis of MS before age 10 years is very rare.^{1–7} The cause of this neurodegenerative disorder is unknown,

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but environmental factors are believed to affect genetically susceptible individuals and increase the likelihood of developing MS. Relapsing-remitting MS (RRMS) is the most common form, and the majority of those diagnosed in childhood have RRMS.^{8,9} Some patients diagnosed with RRMS develop secondary progression later in life. It seems that the younger the patient is at diagnosis, the slower the disease progresses.⁹

Acquired demyelinating syndromes (ADS) presenting as a single demyelinating attack can herald the onset of the chronic MS disorder consisting of recurrent demyelinating attacks in different places within the central

Niðurstöður

- 18 börn og unglíngar
 - 13 eitt stakt kast (Clinically Isolated Syndrome)
 - 2 bráða dreifða heila- og mænubólga (Acute Disseminated Encephalomyelitis)
 - 2 MS í byrjun, 5 fengu greininguna síðar
 - 1 sjóntaugar- og mænubólga (Neuromyelitis Optica)
- Kynjahlutfall jafnt
- Meðalaldur 14 ára
- 60% greind frá janúar – mars
- 5 greind 2009



Faraldsfræði MS í Börnum - MSB

- Nýgengi
 - Erlendar rannsóknir: 0,14 – 0,5/100.000/ár
 - Ísland: 0,45/100.000/ár
- Flestir greinast á unglingsárum
- Um 5% MSF byrjar fyrir 18 ára aldur
- Kynjahlutfall 1:1 fyrir kynþroska

Áhættuþættir MSB

- Lágt D-vítamín
- Hár líkamsþyngdarstuðull (BMI)
- EBV sýking
- HLADRB1
- Sígarettreykur



Meingerð / Orsakir MSB

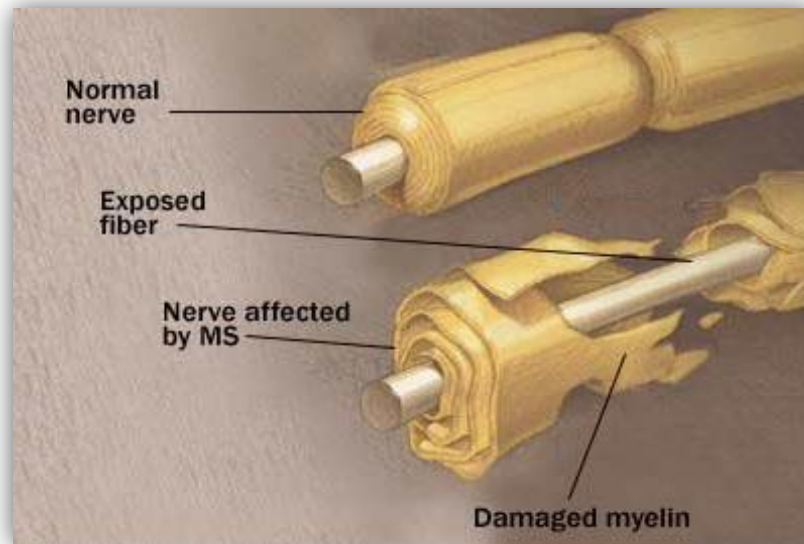
Erfðir
Umhverfi



Bólgublettir
í MTK



Brottfalleinkenni



Upphafseinkenni MSB

- Svipuð og í MSF
- Algengari:
 - Sjóntaugarbólga (Optic Neuritis)
 - Jafnvægistrufnanir (Ataxia)
 - Heilakvilli (Encephalopathy)
- 25% af börnum með ADEM fá MS



Horfur MSB Samanborið við MSF

- 97-99% hafa MS í köstum (RRMS)
- 2 – 3 x fleiri köst á fyrstu 6 árum sjúkdómsins
- Fleiri bólgublettir
- Ná sér fyrr
- Lengur að fá varanlega fötlun á EDSS
- Fötlun á EDSS kemur hins vegar 10 árum fyrr



Hugræn Einkenni í MSB



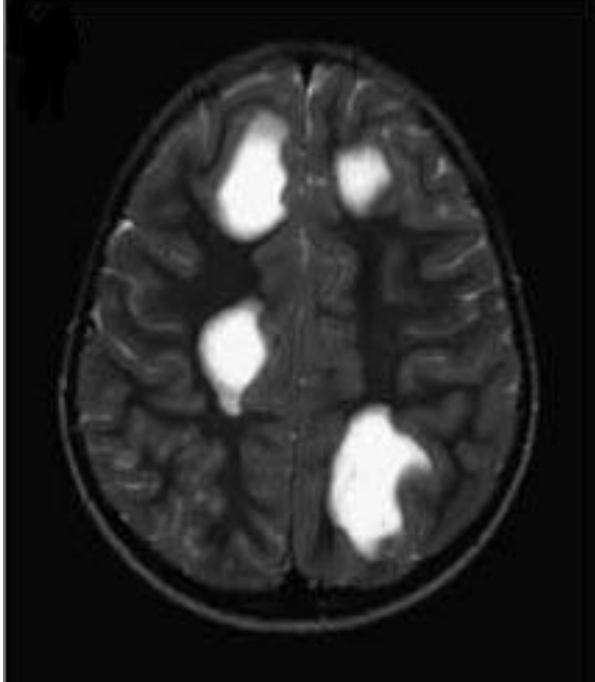
- Þreyta, verkir og hugræn einkenni algeng
- Frávik í
 - Stýringu og skipulagi hugsunar (executive function)
 - Vinnsluhraði
 - Einbeiting
 - Samspil sjónar og handa
- Mikilvægt að fylgjast með þroska og framgangi í námi
- Depurð hjá 20 – 50% með MSB

Hugræn Einkenni í MSB

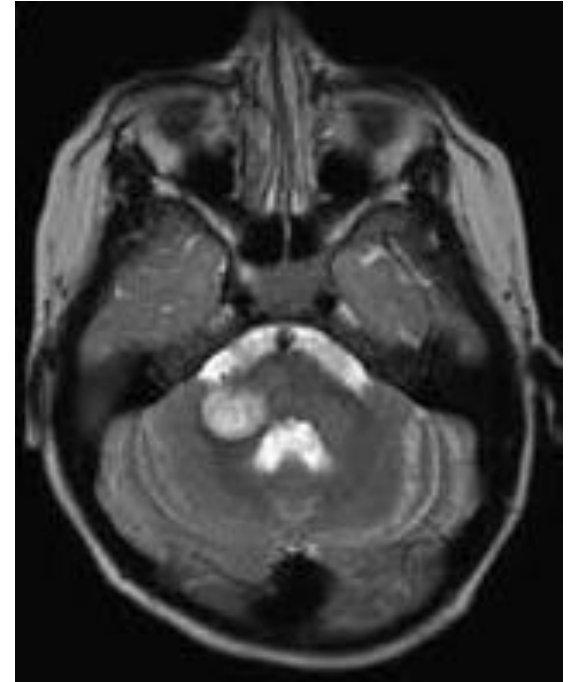
- Óljóst:
 - Hvernig er best að skima
 - Hvað klínísku einkenni eru hagstæð/óhagstæð
 - Á hvaða þroskastigi/aldri heilinn er viðkvæmastur
- Rúmmál heila og höfuðstærð



Mismunargreiningar í MSB

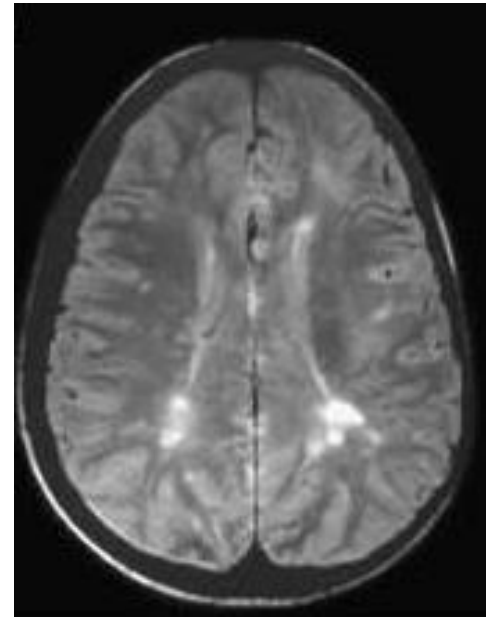


- Sýkingar
- Bráð dreifð heila- og mænubólga (ADEM)
- Efnaskiptasjúkdómar
- Æxli



Greiningarskilmerki McDonalds í MSB

- Áreiðanleg í börnum eldri en 11 ára
- Ónákvæm í yngri börnum
- Klínískt eftirlit og MRI myndataka mikilvæg
- Radiologically Isolated Syndrome (RIS)



Meðferð MSB

- Bráðameðferð eins og MSF
- Fyrirbyggjandi, sjúkdómsheftandi meðferð
 - Interferon beta
 - Glatriamer acetate
 - Dimethyl fumarate
 - Fingolimod
 - Natalizumab
 - Rituximab
- D - vítamín



*Engin ónæmisbreytandi lyf
skráð fyrir börn*



Meðferð MSB

- Svipuð virkni og aukaverkanir og hjá fullorðnum
- Unglingar ≥ 50 kg fá fullorðinsskammt
- Lyf notað í fullum skammti í amk 6 mánuði
- Nálar og sprautur erfiðar hjá unglungum
- Natalizumab hjá þeim sem ekki svara hinum lyfjunum

Meðferð MSB

- Þreyta
 - Amantadine
 - Modafinil
 - Methylphenidate
- Þunglyndi
 - Fluoxetine

Mismunur á MS í Börnum (MSB) og MS í Fullorðnum (MSF) Hugsanlegar Ástæður

- Mismunandi meingerð
- Þroski miðtaugkerfis
- Þroski ónæmiskerfis

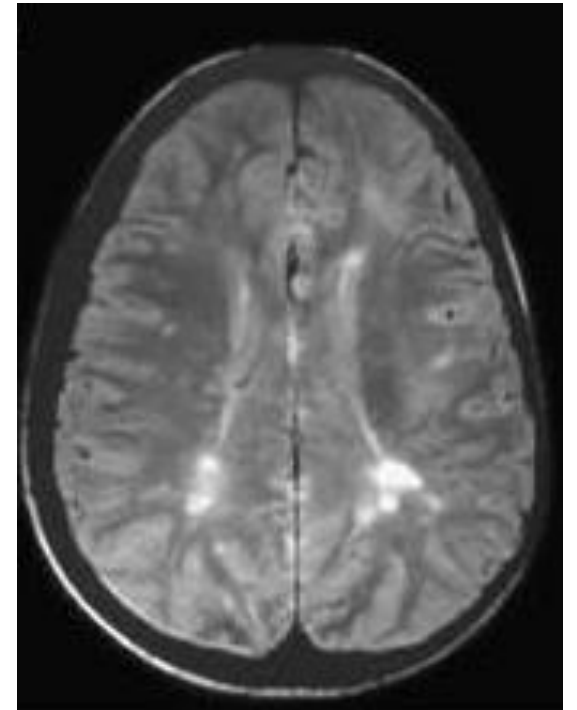


Ónæmiskerfið - MSB

- Sterkari bólgusvar
- Fleiri köst
- Fleiri bólgublettir á MRI
- Einkennin frá mörgum stöðum samtímis
- ADEM

Þroski Miðtaugakerfisins - MSB

- Hvíta efnið þroskast seint
- MS er hvíta efnis sjúkdómur



Er MSB Sami Sjúkdómur og MSF

- Svar: Að hluta
 - Erfða- og áhættuþættir svipaðir
 - Greiningarskilmerki áþekk
 - Virkni meðferðar sambærileg
 - Unglingar með svipaðan sjúkdóm

Helsti munurinn er að sjúkdómurinn leggst á einstaklinga sem eru komnir mislangt á veg í þroska á hvíta efni og ónæmiskerfi