Alder Hey Children's NHS Foundation Trust

Acute demyelination guidelines

Introduction

Please refer to the relevant section in the Oxford Specialist Handbook Paediatric Neurology if available.

Children and young people present with acute demyelinating syndromes (ADS) quite frequently; at least once a month to AHCH. These syndromes are neuroinflammatory disorders of the central nervous syndrome so involve the optic nerve, brain and spine. Early recognition is important as high dose corticosteroid treatment is needed urgently.

This document is to guide the assessment, investigation, and initial management of this group of patients. Patients present with a variety of neurological symptoms and signs depending on which part or parts of the CNS are affected. There are some commoner neurological syndromes that present as an ADS including optic neuritis, encephalopathy often with seizures due to acute disseminated encephalomyelitis (ADEM), myelitis (transverse or longitudinal), ataxia, a brainstem syndrome or a hemiparesis. Some patients may have inflammation in more than one area leading to a polyfocal presentation. Some of these presentations are more common at certain ages for example encephalopathy and seizures in younger children who present with ADEM. With other syndromes generally having a more subacute presentation over days or weeks in older children or teenagers. Infants rarely develop ADS. However, there is a full spectrum of ages of the presentations of this group of disorders. A summary of the different ADS syndromes is shown in the figure below. Rarely, children with Neuromyelitis Optica Spectrum Disorder (NMOSD) may present with gastrointestinal features due to inflammation in the area postrema syndrome of the brain. These children may also have hiccups. Approximately a fifth of patients will have a relapsing course and present with a similar or new neurological syndrome.

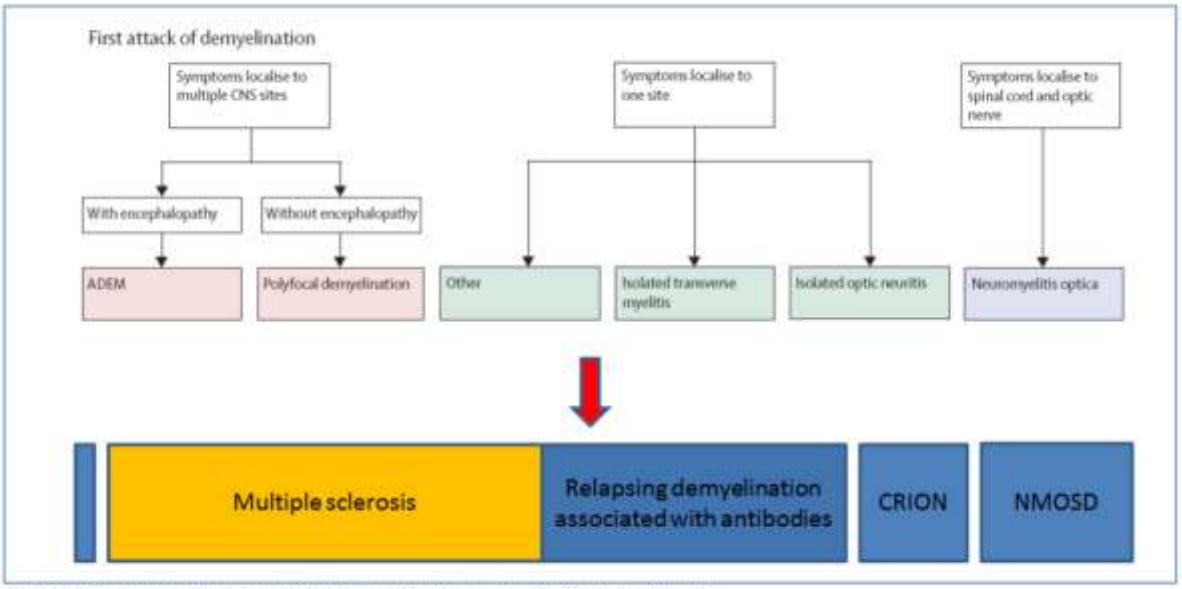

Figure: The spectrum of childhood acquired demyelination syndromes.
ADEM - Acute Disseminated EncephaloMyelitis; CRION - Chronic relapsing inflammatory optic neuritis; NMOSD Neuromyelitis Optica Spectrum Disorder;

Management and Investigation

The acute management for children with ADS starts with a thorough history and examination then MRI scan of the neuro-axis. If ADS is suspected after MRI imaging, the other investigations can be undertaken. These are shown on the figure but for convenience and more detail, they are listed below.

Bloods:

Routine bloods including FBC, renal & liver function.

Send serum for MOG & AQP4 antibodies.

Store a serum sample before start of treatment (essential for potential future investigations)

Vitamin D

IgG for varicella zoster virus (VZV) and Ebstein Barr Virus (EBV).

CSF:

Routine CSF analysis for cell count, culture, Protein, paired serum samples for Glucose and lactate

Oligocloncal bands (requires a paired serum sample)

Store a CSF sample before start of treatment (essential for potential future investigations)

There are some mimics of ADS including CNS infection, certain metabolic disorders e.g. mitochondrial disorders that worsen with fever, systemic inflammatory disorders e.g. SLE, vasculitis, macrophage activation syndromes (HLH), malignancy.

Treatment

All children can be treated with high dose iv methylprednisolone at a dose of 30mg/kg/dose once a day with a maximum dose of 1g od for 3-5 days.

All children diagnosed with ADS need to be discussed with the paediatric neurology team and followed up by the Highly Specialised NHS England commissioned Demyelination Service at AHCH.

The paediatric neurologist on call can help with the decision of how long to give treatment and whether to give an oral taper of prednisolone.

In general, if an oral taper is advised, keep the child on a prednisolone dose of between 5-10mg od, depending on their age, until the antibodies results have become available and the children have been reviewed in the Demyelination clinic.

All children should be treated with a proton pump inhibitor such as omeprazole whilst taking corticosteroid treatment.

Patients and families should be counselled that they must not stop the prednisolone suddenly when they are tapering it off. They must also be informed they are immunocompromised and at risk of infections. Provide a blue steroid card.

They should be given advice to attend their local hospital if the child has a fever of >38.5 degrees centrigrade that has no obvious focus and that does not settle with paracetamol.

In addition, they need to be given advice about what to do if exposed to chicken pox. This information is available in the vaccination Green book. Those with IgG to VZV present, can be considered immune.

If there is a less than full response to iv methylprednisolone, the on call neurologist may advise admitting the child to AHCH for a course of plasma exchange (PLEX).

These children may also need neurorehabilitation services at AHCH.

If a child or young person presents sub-acutely without encephalopathy and the MRI imaging suggests Relapsing Remitting Multiple Sclerosis (RRMS) then please avoid giving this diagnosis to the child and family until they are reviewed in the Demyelination clinic. This diagnosis is for life and has long term implications including psychological distress. There is no indication for starting a disease modifying medication for RRMS acutely. The options will be discussed in the Demyelination clinic.

Discharge

When a child with an ADS is discharged, a clear plan must be given to the family about the risk of immunosuppression, see above. In addition, they must be advised not to receive any live vaccines for 3 months after discontinuing prednisolone. If a child has taken prednisolone for more than 6 weeks, they need to be converted to take hydrocortisone and a LDST needs arranging around 4 months later.

Any child with an ADS can be referred to our CNS Gillian Jones via her AHCH email address gillian.jones@alderhey.nhs.uk. If the child is at AHCH, Gill will try to see them before discharge. She will then help chase up any outstanding investigations and register the child with the Demyelination service run by Dr Rachel Kneen, Consultant Neurologist.

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