

Pediatric Transverse Myelitis

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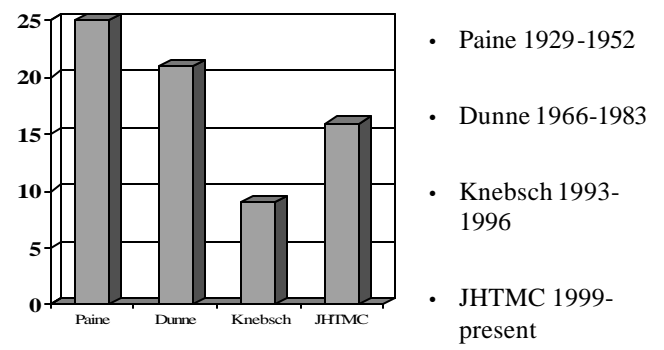
What Makes Pediatric Transverse Myelitis Different?

- Incidence and demographics
- Clinical manifestations
- Etiology
- Diagnosis
- Treatment and prognosis

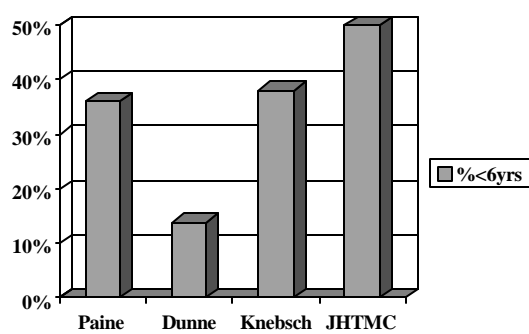
Reported Case Series

- Paine & Byers 1953
- Dunne et al. 1986
- Knebusch et al. 1998
- JHTM center 2001

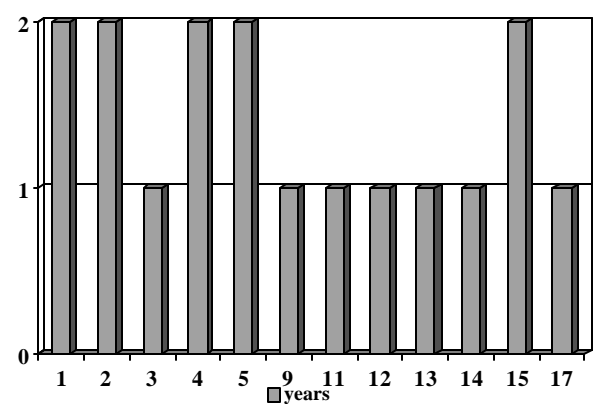
Number of Cases of Pediatric TM



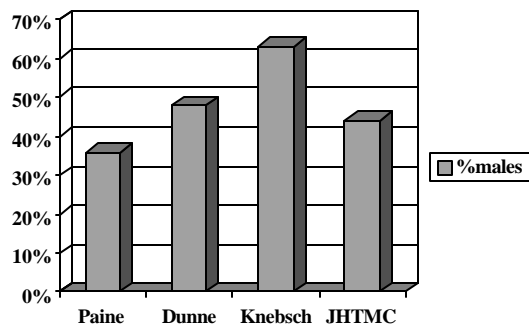
Age of Onset



Age Distribution of JHTMC Series



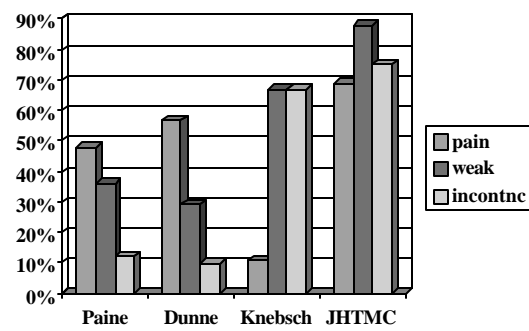
Gender



Clinical Findings

- Sudden onset rapidly progressive weakness of the lower extremities
- Loss of sensation
- Loss of sphincter control
- Pain
- No signs of spinal cord compression or other systemic neurologic disease

Symptoms



Symptoms in Children

- Pain in the back, extremities, or abdomen
 - ◆ Neck stiffness in about 33%
- Fever in about 50%
- Position and vibratory sensation is generally spared if exam reliable

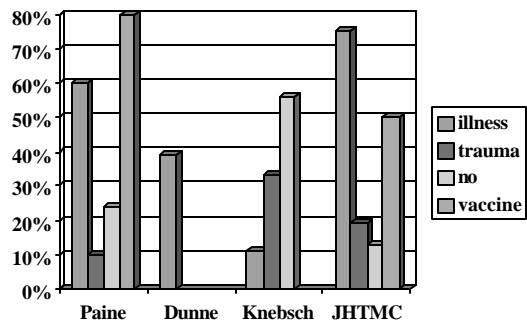
Weakness

- Rapidly progressive paraparesis: usually legs but may sequentially involve legs than the arms
- Flaccid initially with gradually appearing pyramidal signs by the end of the second week of the illness

Sensory Level in Children

- Can be documented in almost all
- Usually T5 and T10
- 20% in cervical
- 10% in lumbar

Etiology



Etiology Factors in Children

- About 2/3 have a history of a recurrent or a concurrent acute infection
- Infectious disease is not associated with the course of the disease

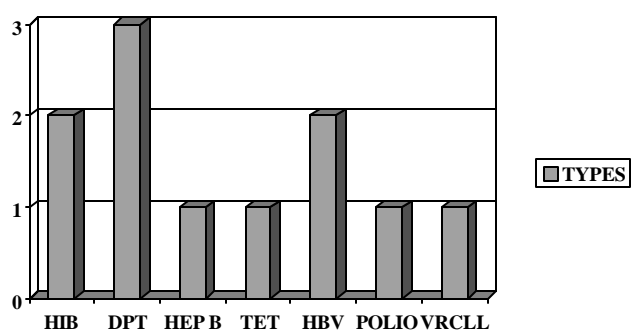
Possible Causative Infectious Agents

- In association or sequel to a large variety of infectious diseases
 - ◆ Herpes
 - ◆ Epstein-Barr virus
 - ◆ Hepatitis B
 - ◆ Influenza
 - ◆ Measles
 - ◆ Mumps
 - ◆ Rubella
 - ◆ Varicella

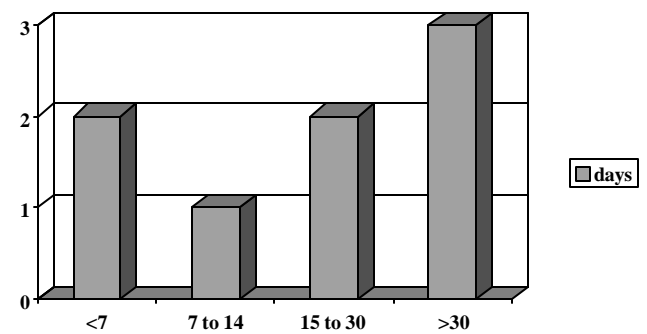
Knebusch – Causative Agent

- 1/8 had rising titers to echovirus 25
- Assumed the others had a para-, post-, or vaccinal related myelitis

Types of Immunization: JHTMC



Time of Preceding Immunization: JHTMC



Etiology

- Suspected cellular autoimmune process
 - ◆ Para- or post-infectious
 - ◆ This should only be diagnosed if other treatable causes can be excluded
- Myelitis from direct infection of the spinal cord
- Myelitis in the context of a systemic autoimmune disease, especially the collagen diseases

Non Parainfectious Causes in Children

- From a medline search for childhood TM from 1990 to 1996
 - ◆ 18 cases infectious
 - ◆ 9 viral; 5 parasitic; 4 bacterial
 - ◆ 2 cases autoimmune disease
 - ◆ 1 SLE; 1 antiphospholipid antibody

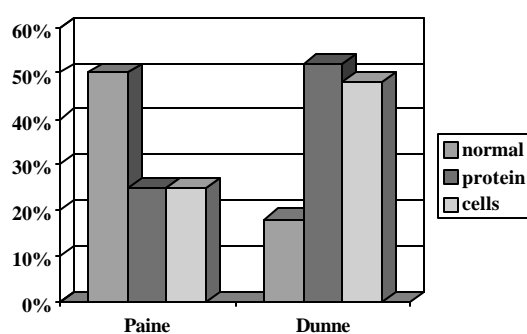
Differential Diagnosis

- Acute infectious polyneuritis
 - ◆ Loss of pain sensation is not as complete as in TM
 - ◆ Proprioception is the most involved sensory modality
 - ◆ Loss of sphincter control and UMN signs favor TM
- Spinal cord space occupying lesions
 - ◆ Neoplasms, epidural abscesses, vascular malformations

Evaluation

- CSF
- EDS
- MRI

CSF Findings



Electrophysiologic Studies

- Helpful in distinguishing central vs. peripheral causes of damage to the CNS
- Altered VEP or BAEP are not compatible with TM
- Prolonged SEP latencies or missing SEP in conjunction with normal sensory nerve action potentials indicate a CNS lesion like TM or MS
- Transcranial magnetic stimulation of limited value in children under 12 due to incomplete maturation of pyramidal tracts

MRI to Rule Out

- Tumor
- Abscess
- Hematoma
- Spinal vascular malformation
- Disc prolapse
- Multiple sclerosis

MRI

- Relatively variable and non-specific
- Used to exclude other lesions that cause paraparesis, especially MS with spinal cord symptoms
- ½ Cases show enlargement of spinal cord on T1
- Unifocal lesions in 80%
- Multisegmental spread is common

Course

- Without therapy
 - ◆ Improvement starts 2 to 12 weeks after the peak of symptoms
- Regaining function as much as 2 years following diagnosis

Treatment

- Exclude other treatable infections that could cause ATM
- Intravenous steroid pulse therapy showed benefit in a pilot study
- Controlled study had not been performed as of 1998

High Dose Methylprednisolone

- 5 children with severe TM
- Open study
- Compared to 10 historical controls
- Outcomes
 - ◆ Median time to walk
 - ◆ Proportion with full recovery
 - ◆ Adverse effects

Sebire et al. Arch Dis Childhood 1997;76:167-168

Treatment

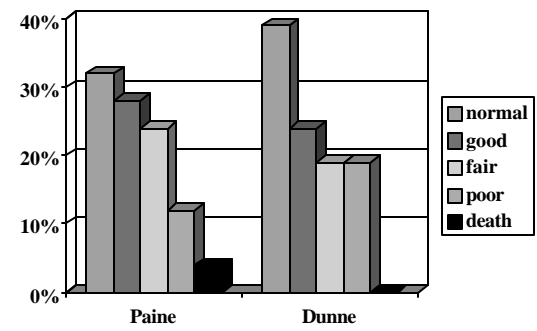
- Knebusch 1998 – unable to determine whether I.V. Pulse steroids lead to improved outcome
- Dunne 1986 – no evidence the outcome was improved in the steroid treated group
- Paine 1953 – one patient treated with I.V. corticotropin followed by I.M. Recovered almost normal function

Outcomes

- Good = gait essentially normal with mild urinary symptoms and/or minimal sensory and upper motor neuron signs: **44%**
- Fair = mild spasticity but independent ambulation, urgency and/or constipation, and some sensory signs: **33%**
- Poor = unable to walk or severe gait disturbance, absence of sphincter control and sensory deficit: **23%**

Knebusch et al. DMCN 1998; 40:631-639

Outcome



Clinical Indicators

- Poor
 - ◆ Backache as the first symptom
 - ◆ Very acute course with maximal symptoms within hours of presentation
 - ◆ Signs of spinal shock
 - ◆ Sensory disturbances up to the cervical level

THANK YOU

