

# Pediatric Transverse Myelitis

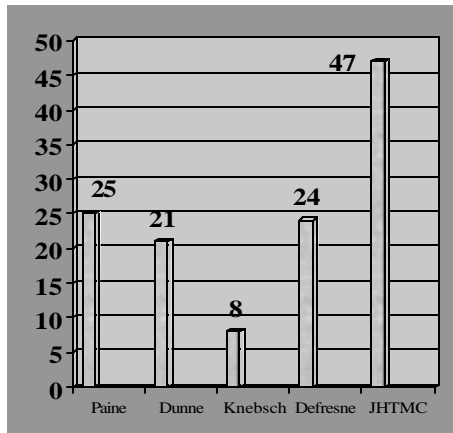
Frank S. Pidcock, M.D.  
Kennedy Krieger Institute  
Johns Hopkins School of Medicine  
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## What do we know about transverse myelitis and children?

- Incidence and demographics
- Clinical manifestations
- Etiology
  - Immunizations
- Functional outcomes & prognosis

## Number of Cases of Pediatric Transverse Myelitis



- Paine 1929-1952
- Dunne 1966-1983
- Knebsch 1993-1996
- Defresne 1965-1995
- JHTMC 2000-present

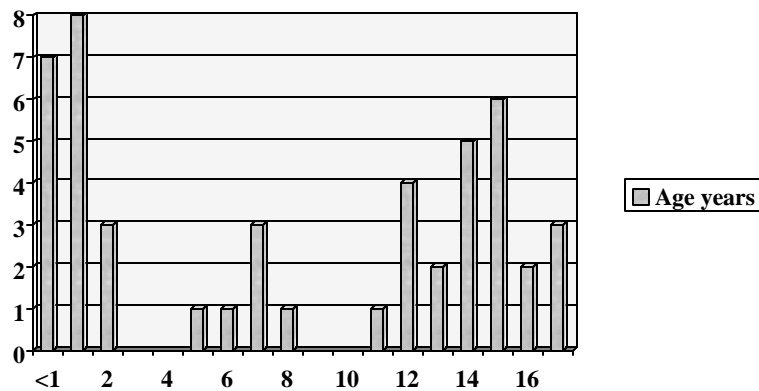
## Johns Hopkins Transverse Myelopathy Center

- Beginning January 2000, patients were entered into a clinical database
- Acute transverse myelitis based on the inclusion criteria and diagnostic algorithm published by the transverse myelitis consortium working group
- Clinical data was collected from retrospective chart review

## Epidemiology-JHTMC

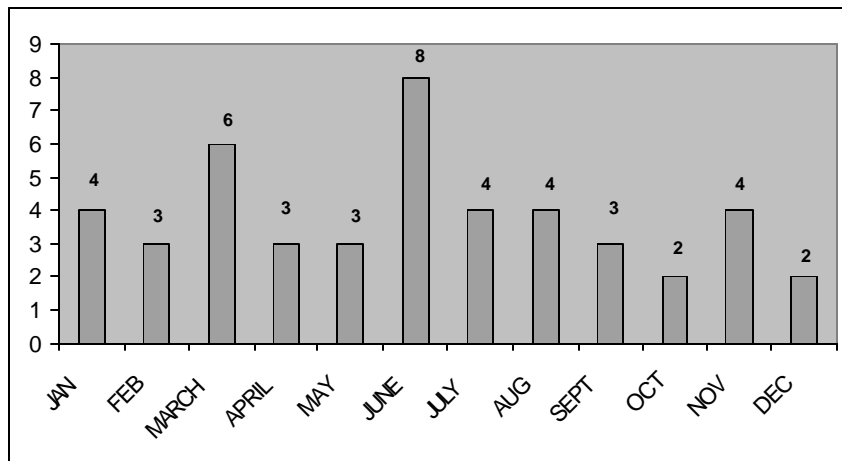
- Forty-seven patients with an acute onset of transverse myelitis at less than 18 years of age
- 89% of the cases were monophasic idiopathic transverse myelitis, two were recurrent cases, and 4 cases has disease associated TM (Devic's, lupus, and ADEM).

## Age of Onset-JHTMC



The average age at acute onset was  $8.3 \pm 0.9$  years

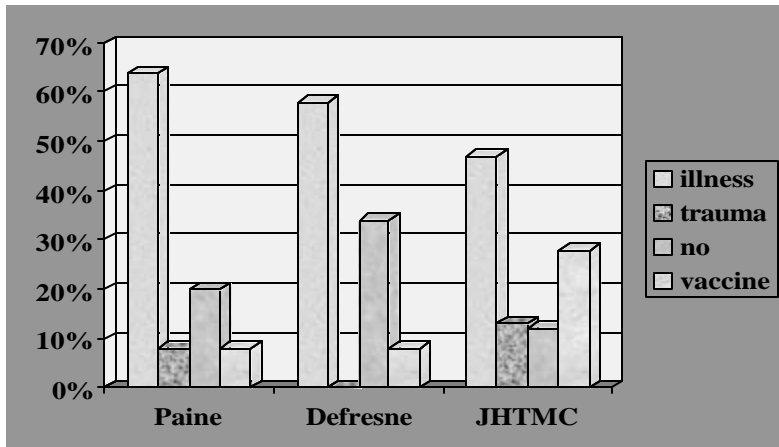
## Distribution of New Cases of Transverse Myelitis By Months



## Immunizations-JHTMC

- 28% reported a preceding immunization (13/47) at an average of  $14 \pm 2$  days prior to the onset of symptoms
- DTP, polio, flu, MMR, small pox vaccine
- All vaccination records were confirmed.

# Etiology



# Currently Recommended Immunization Schedule

Vaccine	Age	Range of live attenuated				CSPV's Immunization				Final vaccine received		
		Birth	1 mo	2 mo	4 mo	6 mo	12 mo	15 mo	18 mo	24 mo	4-6 y	11-12 y
Hepatitis B <sup>1</sup>		HepB 1	HepB 2 (combined HepB+Hib)		HepB 3				HepB series			
Diphtheria, Tetanus, Pertussis <sup>2</sup>			DTaP	DTaP	DTaP		DTaP			DTaP	Td	Td
Meningococcal (Neisseria meningitidis) Type B <sup>3</sup>			Mb	Mb	Mb <sup>4</sup>	Mb						
Inactivated Poliovirus			IPV	IPV	IPV					IPV		
Mening, Mumps, Rubella <sup>4</sup>					MMR 1					MMR 2	MMR 2	
Varicella <sup>5</sup>					Varicella					Varicella		
Pneumococcal <sup>6</sup>			PCV	PCV	PCV	PCV				PCV	PPV	
Hepatitis A <sup>7</sup>										Hepatitis A series		
Influenza <sup>8</sup>					Influenza (yearly)							

<sup>1</sup> Vaccines below this line are for selected populations.

## Illness & Trauma-JHTMC

- 47% (22/47) reported an antecedent illness at an average of  $11 \pm 2$  days prior to the onset of symptoms.
- In 8 cases, antecedent immunization and illness was reported.
- 13% reported antecedent trauma with an average of 8 days prior to the onset of acute neurological symptoms.

## 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

## Clinical Course

- Initial – 5 days: (1-14)
- Plateau - 6 days: (1-26)
- Recovery - regain function as much as 2 years following diagnosis

Defresne et al. 2003

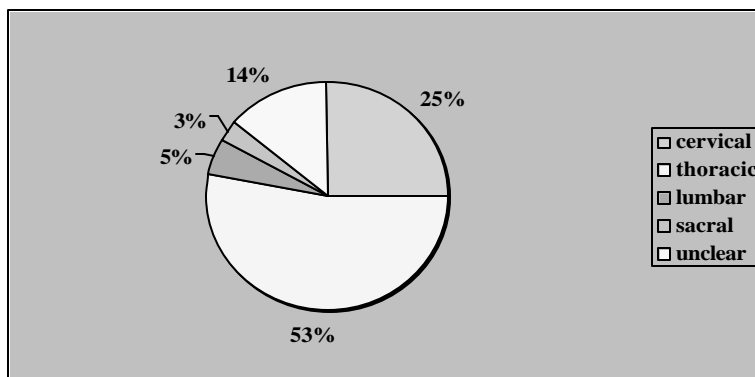
## Acute Symptoms-JHTMC

- Pain 75% (30/40)
- Weakness 89% (42/47)
- Urinary dysfunction 85% (40/47)
- Sensory loss or numbness 91% (40/44)
- At their nadir, 89% (42/47) children were bed bound/wheel chair bound or required assisted ventilation

## Sensory Level in Children

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

## Clinical Sensory Levels in JHTMC Series



## MRI findings- JHTMC series

- T2 cervical – 50%
- T2 thoracic – 40%
- Multifocal – 2%
- T1 hypointense – 38%



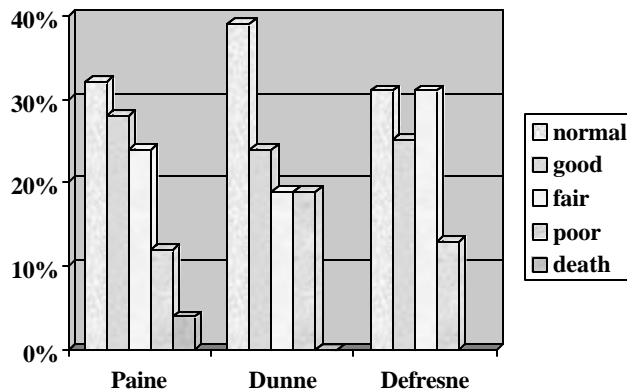
## Functional Outcomes



# Outcomes

- Knebusch
  - 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%**
- Johns Hopkins TM Center
  - Only **19%** experiencing a good outcome by those definitions, likely a reflection of referral bias to a tertiary care center

# Outcome

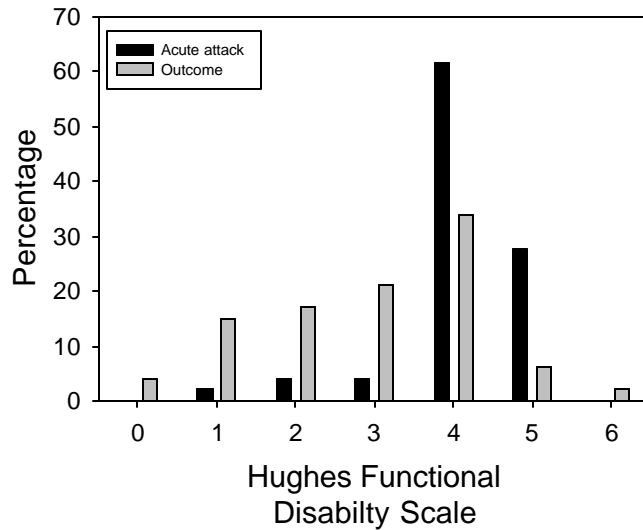


## Outcomes - JHTMC

- Hughes Functional Disability Scale
- WeeFIM
- Evaluated an average of  $8 \pm 1.8$  years (mean  $\pm$  SEM) from the acute onset of disease

## Hughes Functional Disability Scale

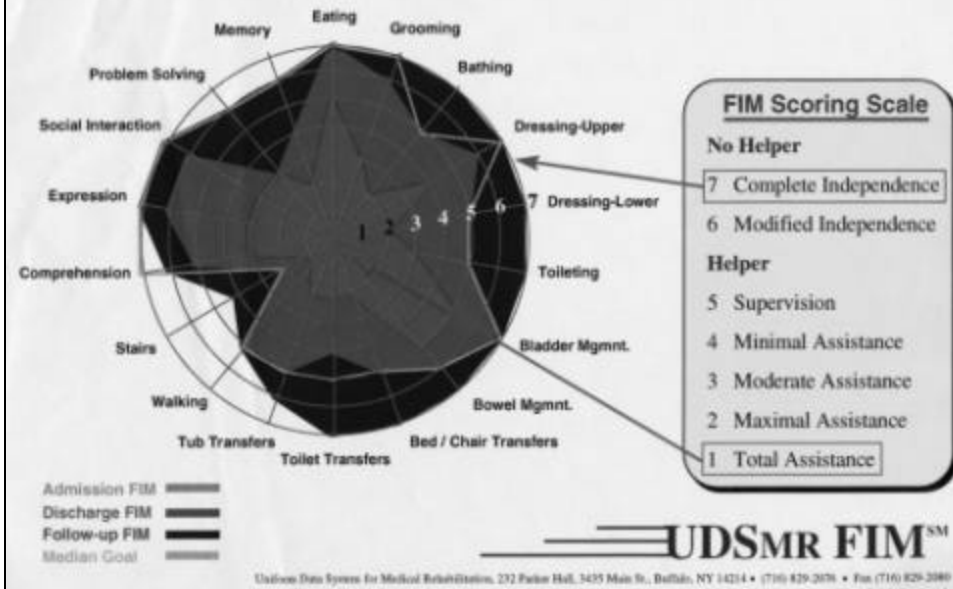
- 0=normal
- 1=minor symptoms, fully capable of manual work
- 2=able to walk more than 30 feet without assistance
- 3=able to walk more than 30 feet with assistance
- 4=bed bound/wheel chair bound
- 5=requires assisted ventilation
- 6= death



## WeeFIM

- An 18 item instrument that measures functional performance in essential daily skills in self-care, mobility and cognitive domains.
- Designed to document functional performance in children and adolescents with either acquired or congenital disabilities.
- Each item rated on a scale from 1-7 (total assistance/dependent to total independence).

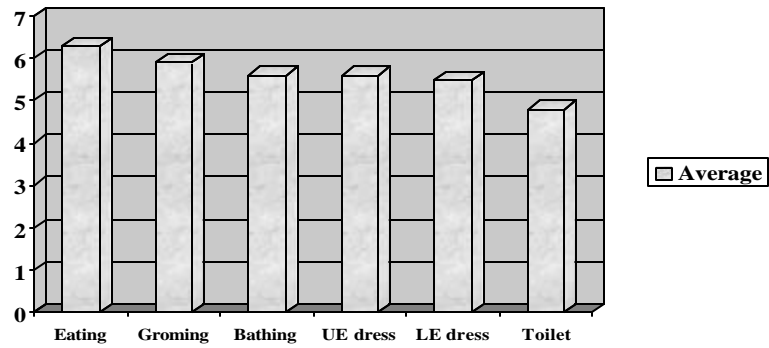
# UDSMR FIM Profile™



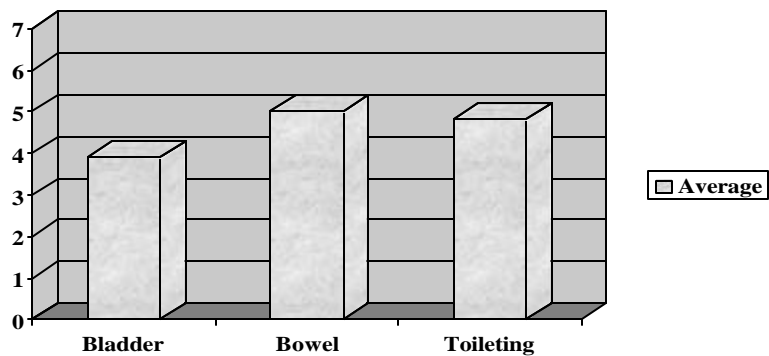
## JHTMC WeeFIM results

Domain	Complete or modified independence % (N)	Modified dependence (moderate assistance to supervision) % (N)	Maximal or total assistance % (N)
Self care	73 (24/33)	12 (4/33)	15 (5/33)
Sphincter control	46 (15/33)	30 (10/33)	34 (8/33)
Mobility (Transfer)	64 (21/33)	18 (6/33)	19 (6/33)
Locomotion	67 (22/33)	30 (10/33)	3 (1/33)
Communication	93 (26/28)	7 (2/28)	-
Social cognition	93 (26/28)	7 (2/28)	-

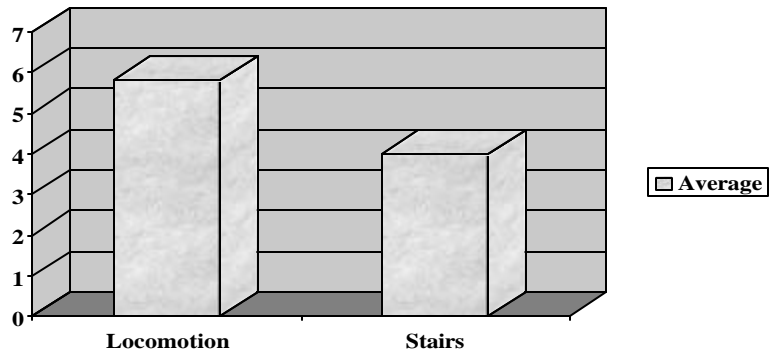
## Average on ADL tasks



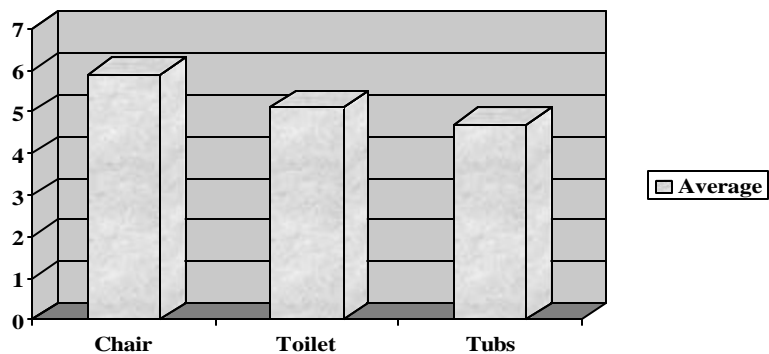
## Averages on Continence



## Average for mobility



## Average for Transfers



## Conclusions

- JHTMC allows more rapid identification of cases
- Probable bias toward more serious cases
- Looking for correlations between interventions and outcomes
- WeeFim scores yield more details on function

**THANK YOU**

