

The Complete Acute and Post-Acute Care Course of Children Affected by Acute Flaccid Myelitis: A Case Series

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Background

Acute flaccid myelitis (AFM)

- Acute onset of asymmetric flaccid weakness in one or more limbs following prodromal viral illness in children.
- Associated with non-polio enteroviruses such as EV-D68 and EV-A71.
- Treatment is mainly supportive.
- Complications include deconditioning, respiratory failure, dysphagia, and impairments in mobility and self-care.
- Dearth of literature describing rehabilitation management and functional outcomes.

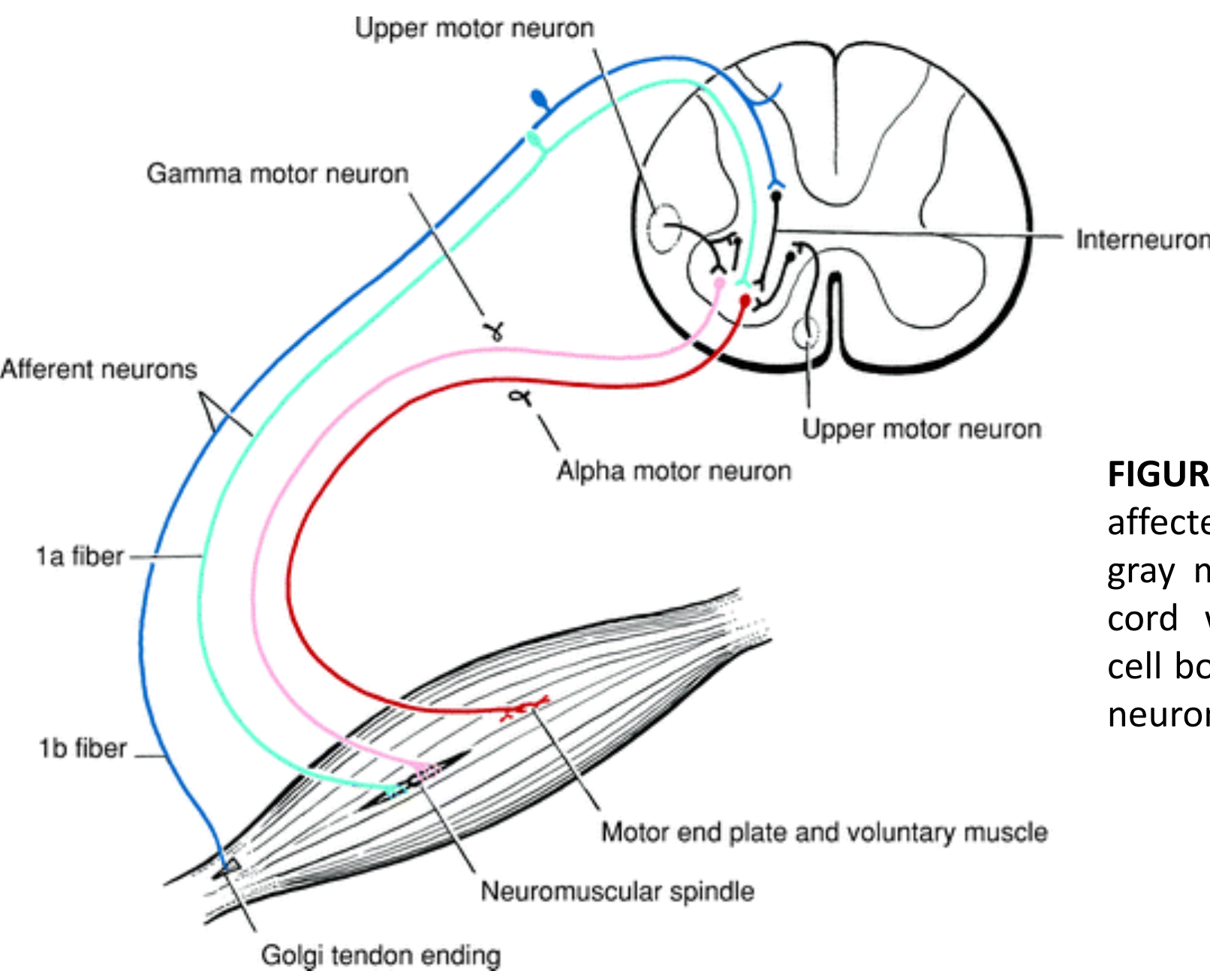


FIGURE 1: The area affected in AFM is the gray matter of the spinal cord which includes the cell bodies of alpha motor neurons.

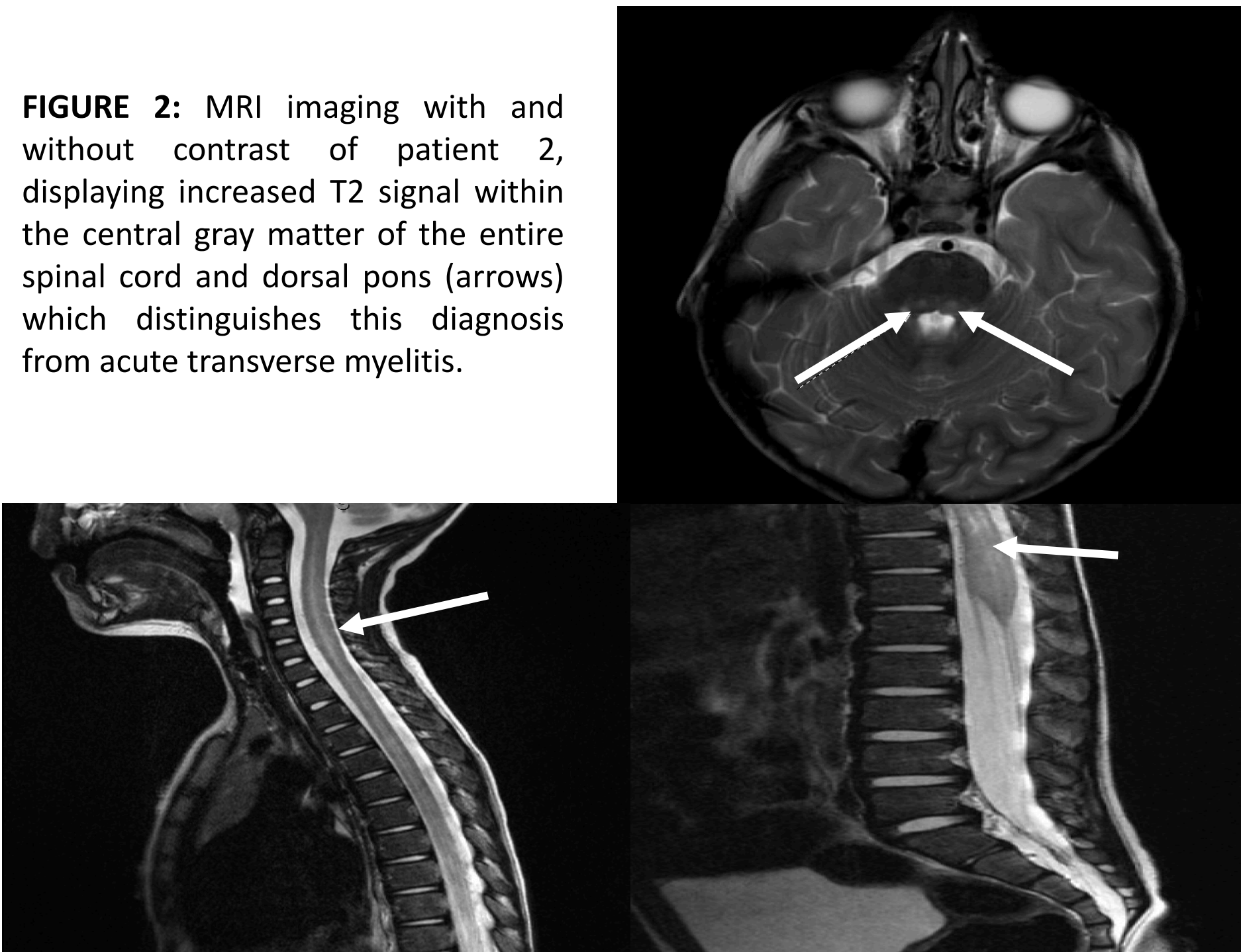
Objectives

To describe the rehabilitation management and functional outcomes in children diagnosed with AFM.

Methods

IRB approved retrospective chart review case series of 6 cases of AFM that occurred at the UPMC Children’s Hospital of Pittsburgh (CHP) between January 2014 and October 2019.

FIGURE 2: MRI imaging with and without contrast of patient 2, displaying increased T2 signal within the central gray matter of the entire spinal cord and dorsal pons (arrows) which distinguishes this diagnosis from acute transverse myelitis.



Results

TABLE 1: Patient demographics and clinical characteristics						
	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6
Age	11 months	2 years, 7 months	3 years, 1 month	3 years, 7 months	3 years, 10 months	6 years, 7 months
Gender	Female	Male	Male	Male	Male	Male
Medical History	Previously healthy	Previously healthy	Previously healthy	ITP	Previously healthy	Motor tics
Prodromal Symptoms	Fever, nausea, vomiting, weakness	Fever, congestion, cough, rhinorrhea	Fever, sore throat, vomiting, weakness	Fever, abdominal pain, vomiting, headache, left upper extremity weakness	Congestion, headaches, decreased appetite, weight loss	Fever, cough, abdominal pain, vomiting, poor appetite, weakness
Clinical Findings	Flaccid quadriplegia, weak cry, diminished or absent reflexes, respiratory distress	Flaccid paraparesis, diminished or absent reflexes, bulbar weakness	Flaccid quadriplegia, diminished reflexes, respiratory distress	Flaccid quadriplegia, right cranial nerve VI palsy, diminished reflexes, respiratory distress	Right lower extremity weakness, absent reflexes in bilateral lower extremities	Bilateral upper extremity weakness, diminished upper extremity reflexes, slurred speech
Brain and spine MRI with and without contrast	Increased T2 signal within the medullary tegmentum, dentate nuclei, and central gray matter of the spinal cord from C2-C6 and T12	Increased T2 signal within the dorsal pons and within the central gray matter of the entire spinal cord	Increased T2 signal within the central gray matter from C1-C7 and T7 -T10, as well as the dorsal pons and midbrain	Increased T2 signal within the posterior medulla, pons, and abnormal signal in the central spinal cord	Increased T2 signal within the anterior cauda equina nerve roots, small syrinx from T11-L1, and mild abnormal T2 signal within distal cord	Increased T2 signal throughout central gray matter of spinal cord, most prominent in anterior horns
Cerebrospinal Fluid	Lymphocytic pleocytosis	Lymphocytic pleocytosis	Lymphocytic pleocytosis	Lymphocytic pleocytosis	Lymphocytic pleocytosis, many RBCs, +EBV PCR	Lymphocytic pleocytosis
Respiratory Viral Panel	+Rhinovirus, +Enterovirus	Negative	+Rhinovirus, +Enterovirus	+Adenovirus C, +Rhinovirus	Negative	Negative
Acute Treatment	Methylprednisolone, IVIG, fluoxetine, ceftriaxone, acyclovir, tracheostomy, gastrostomy tube	Methylprednisolone, IVIG, fluoxetine, ceftriaxone, acyclovir, tracheostomy, gastrostomy tube	Methylprednisolone, IVIG, fluoxetine, ceftriaxone, acyclovir	Methylprednisolone, PLEX, IVIG, amoxicillin, tracheostomy, gastrostomy tube	Methylprednisolone, IVIG	IVIG, fluoxetine, vancomycin, ceftriaxone, acyclovir, Unasyn, tracheostomy, gastrostomy tube
Abbreviations: ITP, immune thrombocytopenic purpura; IVIG, Intravenous immunoglobulin; RBC, red blood cells; PLEX, plasmapheresis						

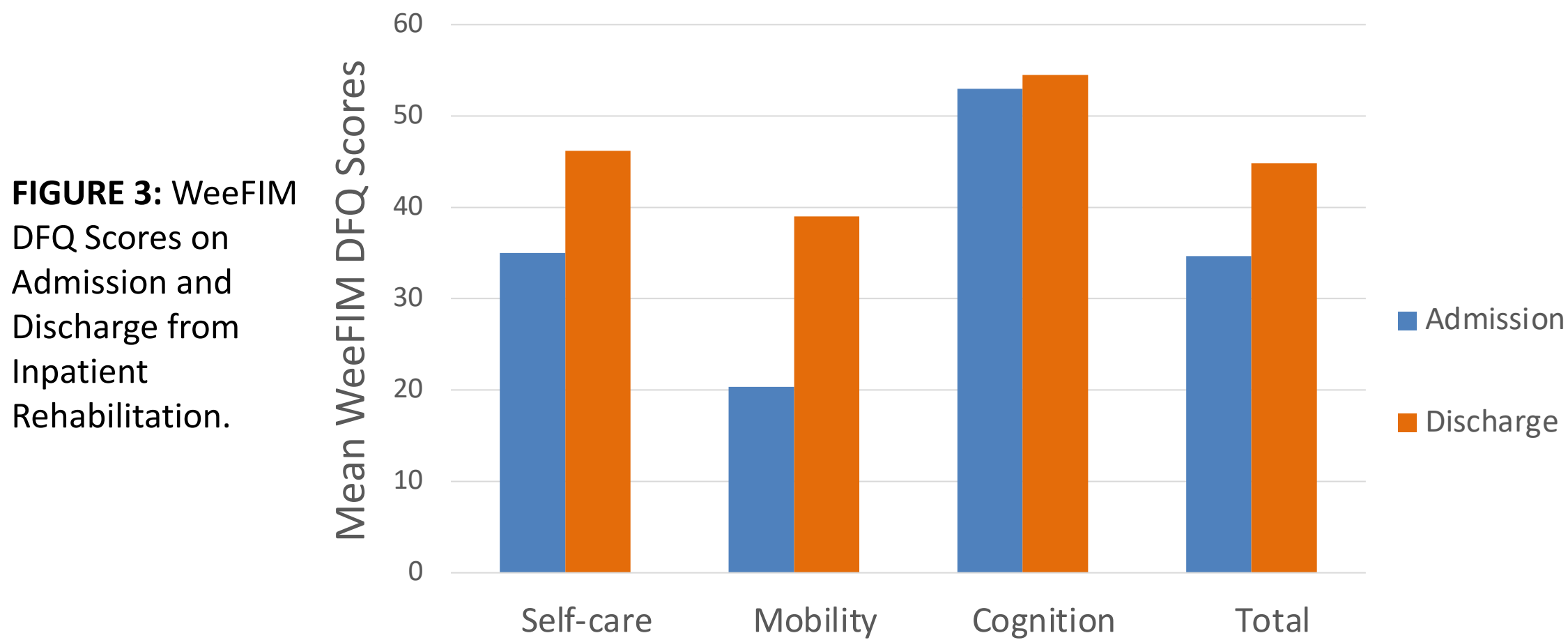


FIGURE 3: WeeFIM DFQ Scores on Admission and Discharge from Inpatient Rehabilitation.

FIGURE 5: Illustration of FES with electrodes placed along the path of the efferent motor nerve and at the muscle of interest. Stimulation patterns vary by current and frequency.

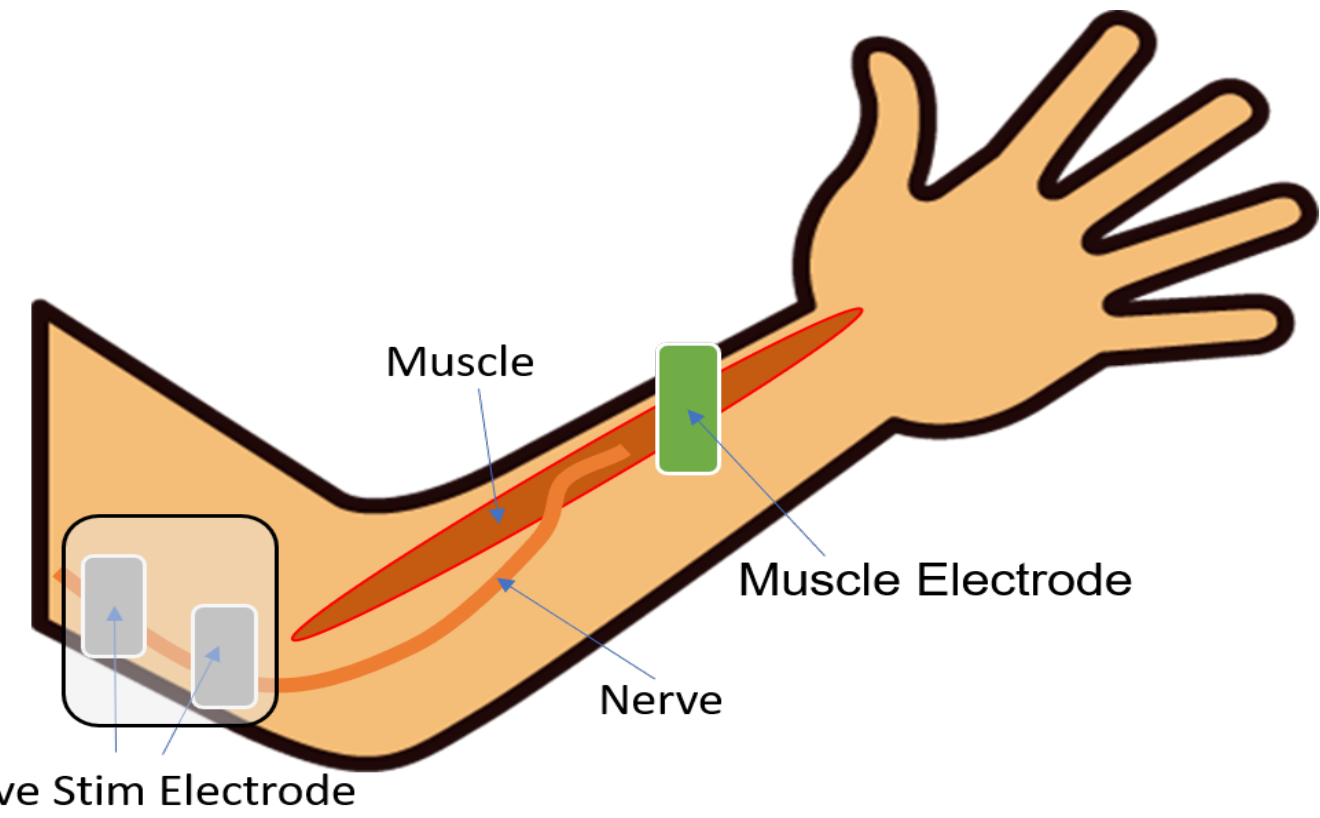


FIGURE 6: Example illustration of nerve transfer performed between the radial nerve (RN) and the median nerve (MN).

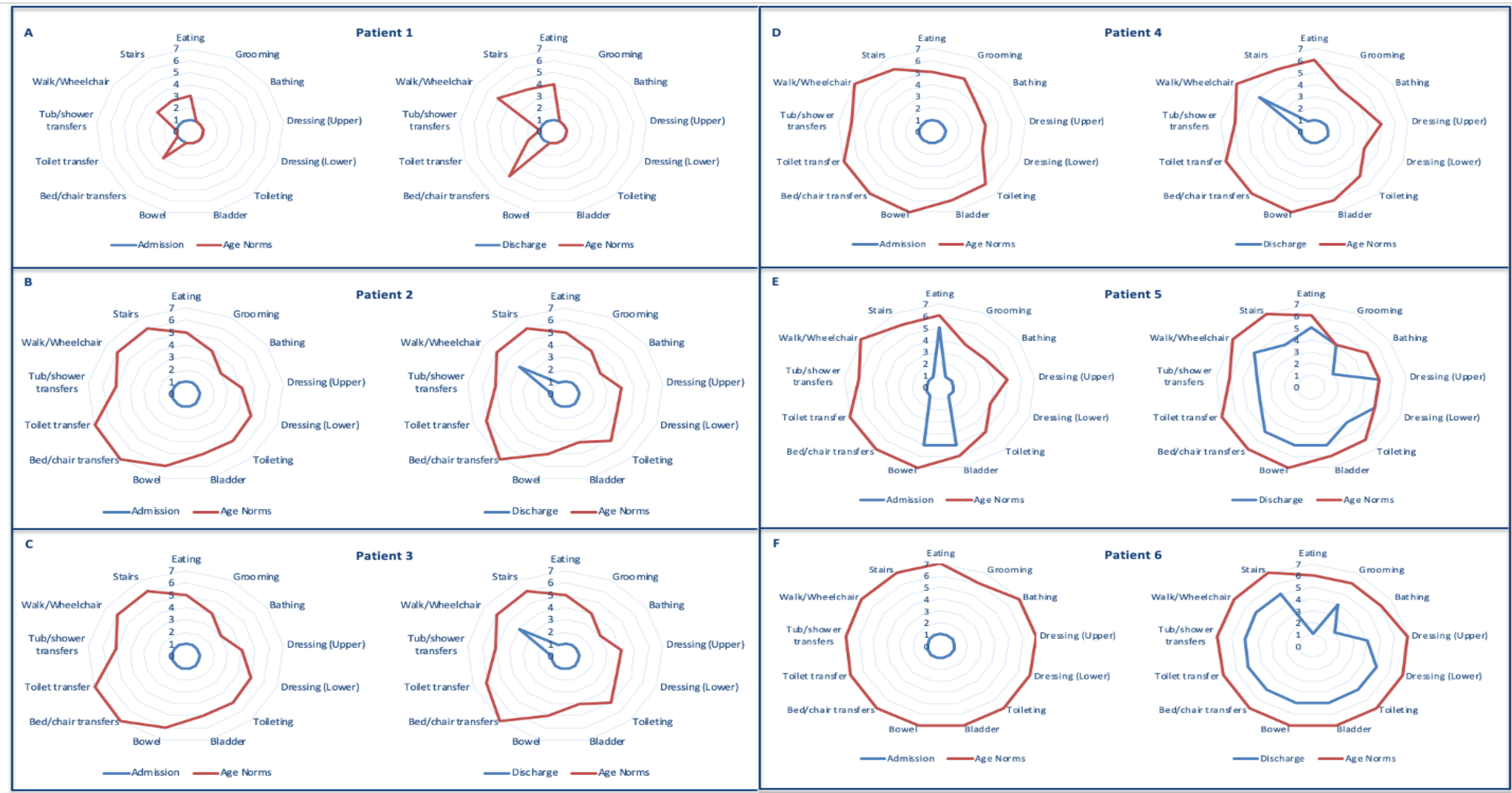
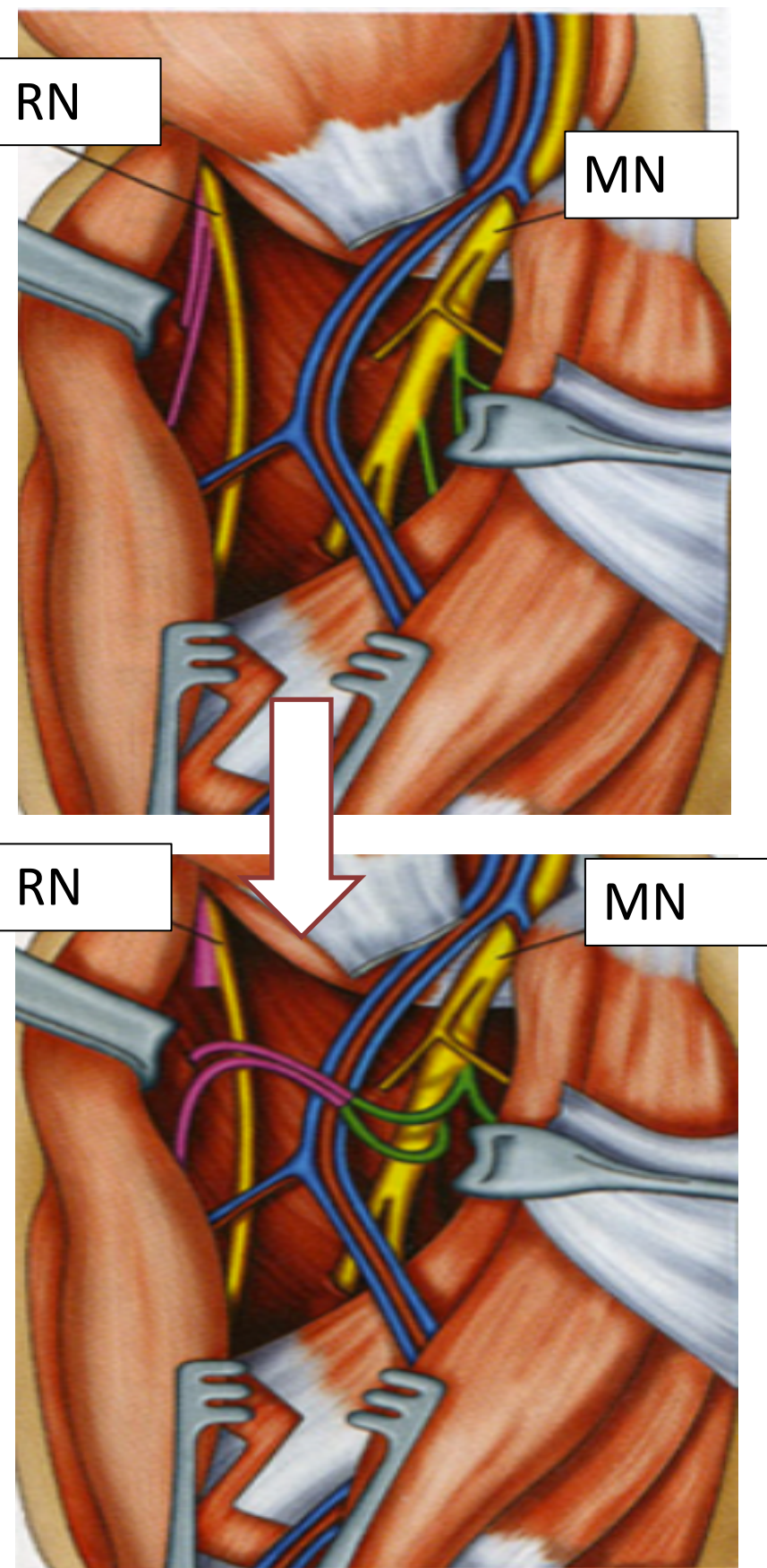


FIGURE 4: WeeFIM self-care and mobility scores at admission and discharge (blue) from comprehensive inpatient rehabilitation. WeeFIM scores for each patient are compared to age norms (orange).

Discussion

Acute Care Highlights

- 6 children** (5 boys and 1 girl; average age 3.4 years) diagnosed with AFM had imaging findings showing gray matter involvement in brainstem as well as the cervical spinal cord (4 cases) and thoraco-lumbar spinal cord (2 cases).
- All 6 patients received **methylprednisolone** and **IVIG** and 1 patient received **PLEX**.
 - Recent studies in mice indicate that early steroids may be harmful.
- Complications included respiratory failure requiring tracheostomy, dysphagia requiring gastrostomy, and respiratory infection.

Post-Acute Care Highlights

- 4 patients** demonstrated improvement in self-care, mobility, and total WeeFIM developmental functional quotient (DFQ) scores.
 - Average total WeeFIM DFQ was 34.7 on admission, 44.8 at discharge, and functional status remained below age-expectation.
 - All patients were discharged home with appropriate equipment, orthotics, and therapies.
 - 3 children** required mechanical ventilation and **4 children** were wheelchair dependent at discharge.
- FES** facilitated muscle contraction in 3 patients but was limited by pain and fear avoidance.
- Nerve transfer** surgeries were performed on 3 children after discharge (average 14.3 months after presentation, range 11-18 months).

Conclusions

- Comprehensive inpatient rehabilitation is critical to maximize functional outcomes and quality of life for patients with AFM.
- Patients made functional gains in different aspects of age-appropriate mobility, self-care, feeding, and communication.

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