

5th Annual Conference of the Periodic Paralysis Association

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Las Vegas, Nevada

EMS Orientation to the Periodic Paralyse

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Association

Unique Opportunity

- Thank you for your interest and support.

What are the Periodic Paralyses and Non-Dystrophic Myotonias?

NORMAL PHYSIOLOGY

EACH DISORDER

FIELD MANAGEMENT

Q & A

Normal Physiology

- Nerves = electric wires
- Transmit signals from
BRAIN → MUSCLE

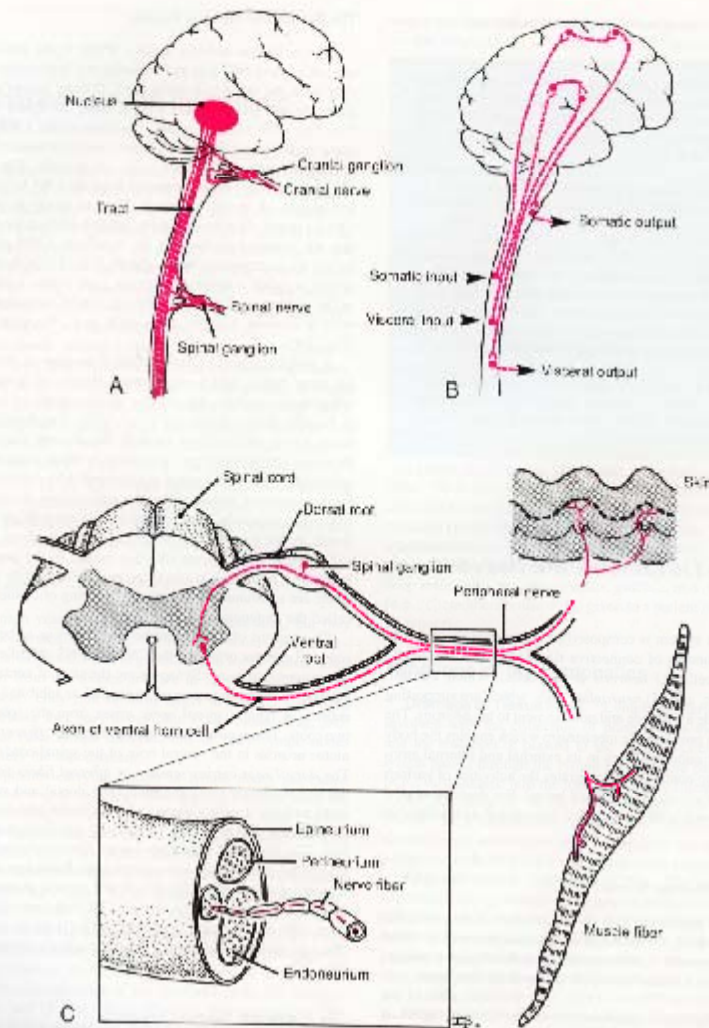


Figure 25. The basic organization of the nervous system. A, Shows a gross tract: a collection of nerve fibers arising from a nucleus (a collection of nerve cells) in the brain and passing into the spinal cord. The cranial and spinal nerves arise from the brain and spinal cord,

respectively. B, Somatic and visceral sensory input on the right side and somatic and autonomic output on the left side. C, The organization of the peripheral nervous system (PNS).

Table 4.
Functions of
Different Organs
of the
Nervous System

Brain
Spinal cord
Sensory
Motor
Autonomic
Visceral
Sensory
Motor

Ciliary muscles

Conjunctiva
Sweat glands
Lacrimal glands
Salivary glands
Gastric glands
Intestinal glands
Suprarenal gland
Cortex
Liver
Spleen
Lactiferous glands
Pancreas
Spleen
Nails

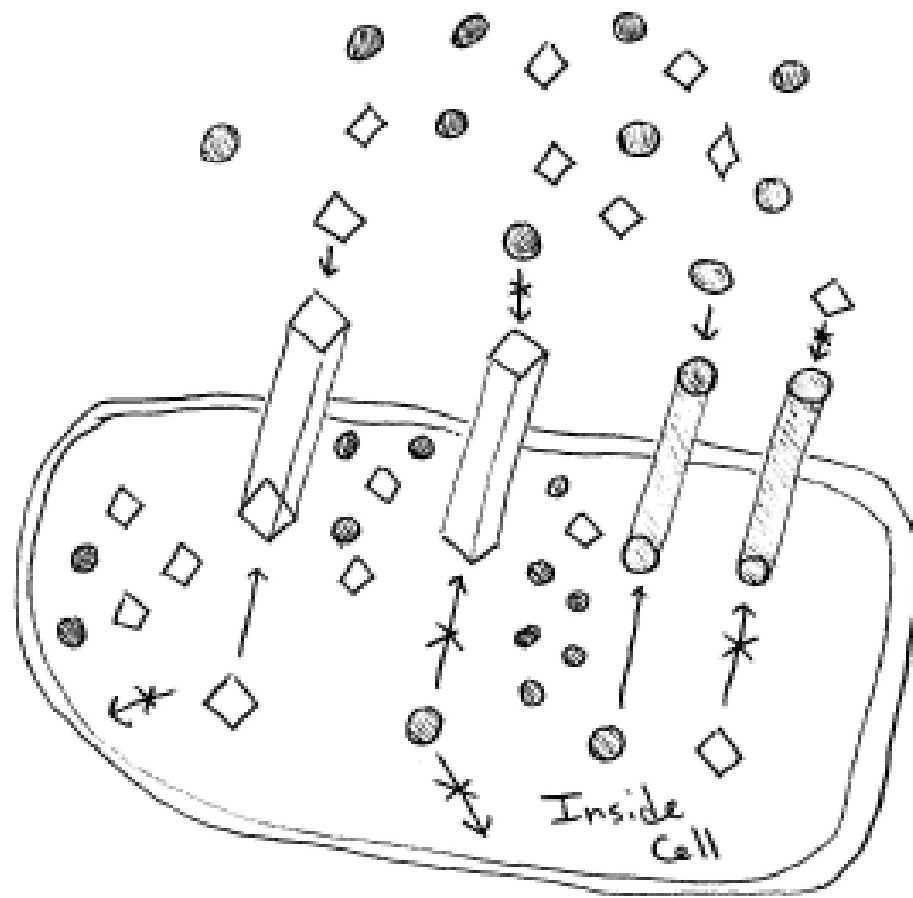
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More Normal Physiology

Muscle cells

- potassium & sodium
- inside and outside of cell membranes

● = Potassium (K^+)
◊ = Sodium (Na^+)



Outside cell

Inside Cell

More Normal Physiology

Muscle cells respond to nerve signals:

- ions change location around cell membrane:

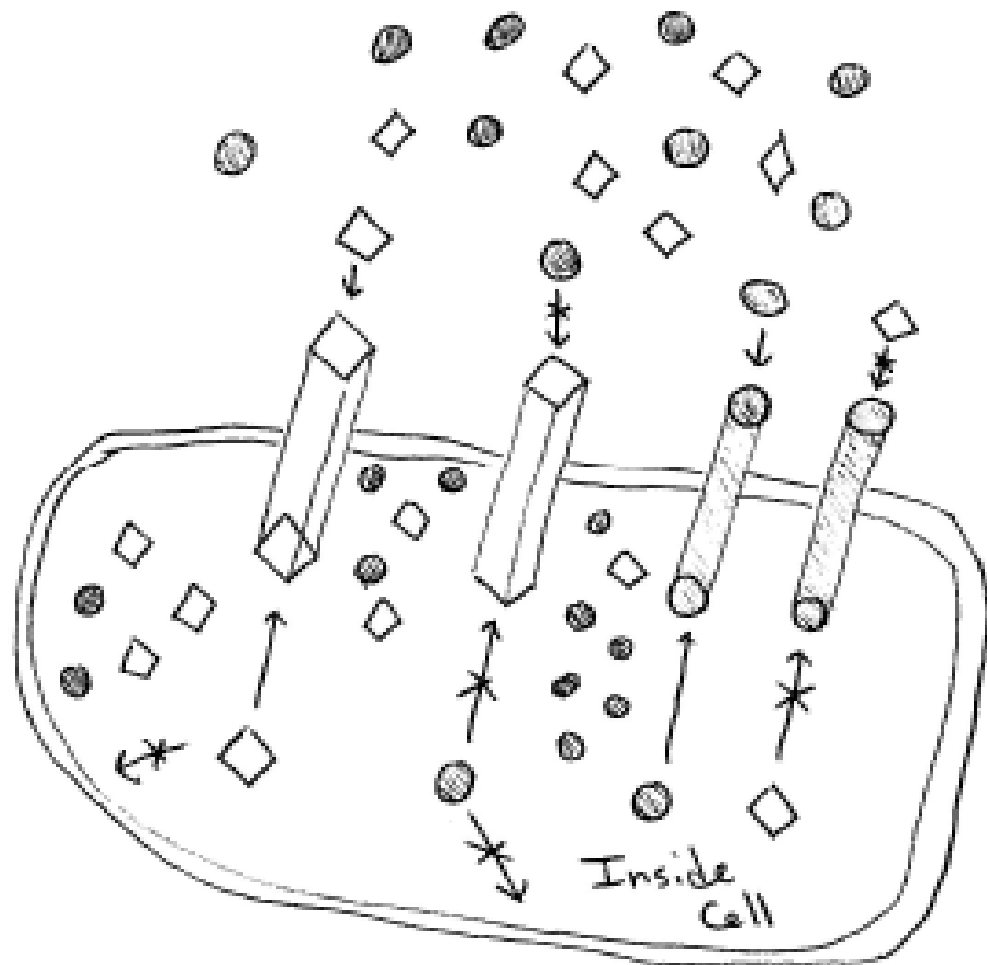
Inside → outside

Outside → inside

More Normal Physiology

- The ions cross the muscle membrane through selective ION CHANNELS.
 - Na^+ → sodium channel
 - K^+ → potassium channel
 - Ca^{++} → calcium channel
 - Cl^- → chloride channel

● = Potassium (K^+)
◊ = Sodium (Na^+)



Outside cell

Defect in Periodic Paralysis

- Periodic paralysis & the non-dystrophic myotonias
 - muscle weakness and/or muscle stiffness
- Because: ION CHANNELS in the muscle membrane are faulty.

Defect in Periodic Paralysis

- The faulty ion channels prevent the potassium and sodium from being where they need to be.
- The misplaced ions cause the ion channels either:
 - to **shut off** inappropriately, causing **weakness**
 - **not to shut off** when they should, causing **stiffness**.

What is meant by weakness

- Sometimes, can move against gravity
- Oftentimes, DEAD WEIGHT
 - Most often extremities (arms, legs)
- Usually, respiratory muscles intact
- Occasionally, cannot speak or blink

- **MIMICS PARALYSIS**

One name, Multiple types

- **Hypokalemic PP**
 - Weakness in setting of LOW serum [K⁺]
- **Hyperkalemic PP**
 - Weakness in setting of HIGH serum [K⁺]
- **Paramyotonia Congenita**
 - Muscle stiffness, worse with exercise
- **Myotonia Congenita**
 - Muscle stiffness, better with exercise
- **Potassium-Sensitive Myotonia**
 - Muscle stiffness, worsened by K⁺
- **Andersen-Tawil Syndrome (aka Andersen's)**
 - Periodic paralysis, arrhythmia, skeletal defects

Difficulty with Classification

- Overlapping symptoms
 - Example: paralysis AND myotonia
- Wide variety of less characterized signs/symptoms
 - Irritability, subjective SOB, muscle pains, sudden collapse, feeling cold, impaired cognition, myoclonus
- Many people do not fit the exact mold of the classifications below
 - Example: weakness with normal serum [K+], but improves with [K+] intake

Hypokalemic Periodic Paralysis

Familial (Genetic)

- Sodium channel, Potassium Channel, Calcium channel

Secondary (not genetic)

Thyroid over-activity

Endocrine or renal problems

Certain substances and medications

Hypokalemic Periodic Paralysis

Main Symptoms:

weakness

low serum potassium

permanent muscle weakness

Main Triggers:

– Carbohydrates

– Salt

– Rest after exercise

– Stress

– Cold

– Insulin

Hypokalemic Periodic Paralysis

Things that Alleviate Attacks Acutely:

potassium

Things that Prevent Attacks Chronically:

Acetazolamide, dichlorphenamide

Spiroinolactone, triamterene, amiloride

Proper diet, avoidance of triggers

Hyperkalemic Periodic Paralysis

Familial (Genetic): Sodium channel

Secondary: Renal problems

Main Symptoms:

weakness, usually less duration than in hypokalemic periodic paralysis

myotonia (muscle stiffness that can be alleviated by exercise)

high serum potassium

permanent muscle weakness

Hyperkalemic Periodic Paralysis

- Main Triggers:
 - Potassium intake
 - Rest after exercise
 - Stress
 - Hunger
 - Cold
- Things that Alleviate Attacks Acutely:
 - Carbohydrates (simple sugars)
 - Insulin
 - Albuterol
 - Calcium injection

Hyperkalemic Periodic Paralysis

- Things that Prevent Attacks Chronically:
 - acetazolamide, dichlorphenamide
 - other potassium-wasting diuretics (e.g. hydrochlorothiazide)
 - proper diet
 - avoidance of triggers

Andersen-Tawil Syndrome

- Familial (Genetic): potassium channel
- Definition:
 - Periodic Paralysis (hyper or hypo)
 - long QT syndrome (or other arrhythmias)
 - Skeletal abnormalities (wide spaced eyes, low-set ears, webbed fingers or toes)

Andersen-Tawil Syndrome

- Main Symptoms:
 - Weakness
 - Palpitations
 - Atypical attacks with muscle twitching (myoclonus)
 - May mimic seizure
 - Low or high serum potassium (depends on associated periodic paralysis)
- Main Triggers: same as that for the associated type of periodic paralysis
- Things that Alleviate Attacks: same as that for the associated type of periodic paralysis
- Things that Prevent Attacks Chronically: same as that for the associated type of periodic paralysis

Paramyotonia Congenita

Familial (Genetic): sodium channel

Definition: muscle stiffness that worsens with exercise (paradoxical myotonia) and with cold

Main Symptoms:

- Muscle stiffness followed by weakness, especially with exercise in cold weather
- Can be associated with hyperkalemic periodic paralysis
- Normal serum potassium
- No warm-up phenomenon

Paramyotonia Congenita

- Main Triggers:
 - Same as those for hyperkalemic periodic paralysis
 - Cooling and heavy muscular work
- Things that Alleviate Attacks Acutely:
 - Warm environment relieves stiffness
 - No therapy to relieve weakness acutely
 - Same as for hyperkalemic periodic paralysis (if hyperkalemic periodic paralysis is a feature)
- Things that Prevent Attacks Chronically:
 - Acetazolamide
 - Mexilitene
 - Same as for hyperkalemic periodic paralysis if that is a feature

Potassium-Sensitive Myotonia

- Familial (Genetic): Sodium channel
- Main Symptoms:
 - Intermittent, generalized muscle stiffness
 - No weakness
 - Not worsened by cold
- Main Triggers:
 - Potassium ingestion
 - Rest after exercise
 - Succinylcholine anesthesia

Potassium-Sensitive Myotonia

- Things that Alleviate Attacks Acutely:
same as for hyperkalemic periodic paralysis
- Things that Prevent Attacks Chronically:
 - Mexilitene
 - Acetazolamide

Myotonia Congenita

Familial (Genetic): Chloride channel

- Main Symptoms: muscle stiffness
- Main Triggers: sudden exercise, sudden noise
- Things that Alleviate Attacks Acutely: repeated movement of stiff muscle (warm-up phenomenon)
- Things that Prevent Attacks Chronically: mexilitene, acetazolamide, phenytoin, quinine, carbamazepine

Management in the Field

Sunrise Hospital

- **Address:** 3186 South Maryland Parkway, Las Vegas, Nevada 89109,
- **Telephone:** (702) 731-8000
- **Fax:** (702) 731-8668
- **ER aware** of possible periodic paralysis patients
- **Dr. Alfreda Maller** is on call for neurology and will be available to manage cases.

Scenario

- Acute Attack
- Patient's buddy is usually present
- Best case: Patient (and/or buddy) can speak and explain the situation
- Worst case: Patient is alone, cannot speak
- Usually EMS are called for the **Andersen** attacks or for severe attacks of **Hypokalemic Periodic Paralysis**

Establish Preliminary Diagnosis: Listen to the Patient and Friends

- Patients attending this conference are likely to have some form of periodic paralysis.
- Patients typically carry a valid diagnosis.
- Patients typically know what to do.

Patients can present with:

- a. Weakness of extremities
- b. Myoclonic jerks with inability to speak, mimicking seizure activity
- c. Typically have normal tactile and auditory sensation even though they may not be able to speak
- d. Arrhythmias
- e. Asthma or Diabetes complicating potassium management
- f. Extreme muscle stiffness, sometimes painful, in some muscles

What to do First

- **Airway, Breathing, Circulation**
 - Often intact
- Risk of **aspiration**
- **EKG** for rhythm

NON-ACTION is ACTION

- Get history
- Patient is often very particular
 - **how they are moved**
 - **Position**
- Respect patient's **frustration of powerlessness**

Initial Assessment

- What are we dealing with:
 - a) **low serum potassium** with attack (**hypoKPP**),
 - b) **high serum potassium** with attack (**hyperKPP**)
 - c) muscle stiffness (usu. with high K^+).
- **Patients get into worst problems if their normal routine of self-medication has not alleviated attacks.**
- This requires hospital monitoring.

**Gray Area:
Transport Patient to ER or
Leave Them Be**

- Take patient to Sunrise Hospital:
 - When in doubt
 - Upon patient request
 - Respiratory or cardiovascular instability

Quandary:
Patient Wants to Stay,
EMS Wants ER

- Not an easy decision
- **Please respect patient's right to refuse treatment**

Patient's Perspective

- Patient may feel they are able to manage.
- Patients often know when they are in over their head.
- From experience, they know when to watch and wait vs. when to go to the hospital.
- Oftentimes, they appear worse off than they are.
- Sometimes EMS is called by someone other than the patient before the patient has self-medicated.
- Patients typically know what medicines and what doses of medicine (especially potassium) help them.
- Patients typically know the expected duration of their attacks.

More Patient's Perspective

- Generally, the patients will come out of the attack within minutes to hours.
- Patience is imperative.
- Time must be allowed for medications to begin working.

EMS Perspective

- **Medicolegal** issues make it difficult to leave the patient in paralyzed condition
- Patients often try to be “**heros**” and don’t want to ruin an activity for their companions by an ER visit
- **Cannot be sure** of situation without blood tests (esp. serum potassium)

Management in the Field

- As in any emergency situation, **Airway, Breathing, and Circulation** are priority.
- **EKG for rhythm** is appropriate.
- Appreciate **risk for aspiration** (but usually not occur) BUT...patients are usually most comfortable on their back
- **What other routine procedures do you do?**

Management in the Field

- If possible, a **red-top** tube drawn for serum electrolyte (**SMA 7**) would be helpful to the ER staff prior to patient arrival (however, **prompt processing** is needed to assure correct potassium measurement).
- **DO NOT:** administer *I.V. dextrose or saline* (as these are potential **triggers** for periodic **paralysis**) unless indicated for supervening medical conditions

Fluid Management

- Intravenous Fluids are typically not necessary.
- If intravenous fluids are necessary, understand that the glucose in D5W and the sodium in 1/2 NS can **worsen hypokalemia** in hypokalemic periodic paralysis

Potassium Management (for HypoKPP and Andersen)

- Electrolyte management can be **done in the ER** and is best reserved for after a **baseline serum potassium** can be established. (Decision to give potassium by mouth on site must be made together with the patient.)
- Oral potassium replacement (via KCl powder or effervescent tablet dissolved in water) is favored over intravenous replacement unless the patient cannot take medication orally.

Take Buddy to ER

- If at all possible
- Compensates for ER neglect since likely patient will be stable

Special Cases

- If a patient is asthmatic and has Hypokalemic Periodic Paralysis, understand that albuterol will worsen hypokalemia and weakness but may be necessary for bronchodilation.
- If patient is diabetic, insulin worsens hypokalemia and induces paralysis but may be necessary for glucose control.

Medications You May Encounter

- Acetazolamide, dichlorphenamide (All)
 - Carbonic Anhydrase Inhibitors, potassium wasting diuretics, effective for many forms of periodic paralysis
- Spironolactone, triamterene (HypoKPP)
 - Potassium-sparing diuretics
- Hydrochlorothiazide (HyperKPP)
 - Potassium-wasting diuretic

Medications

- Potassium chloride
 - (patient likely will have taken some dose prior to EMS arrival)
 - Effervescent tablets
 - Powder
 - Sustained Release pills
- Calcium and magnesium supplements

**Thank You for your
kind attention!!!**