Lance-Adams Syndrome: Effective Treatment and Functional Improvement with Phenytoin  

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Abstract

Post-hypoxic myoclonus (PHM), or Lance-Adams Syndrome, is a rare and difficult to treat condition that is characterized by involuntary muscular jerking movements in people who have sustained a cerebral event. Both the incidence and pathogenesis have not clearly been established, and little is known about effective treatments for this condition. This is a rare syndrome, and the successful treatment for PHM has been limited. This report is unusual in that it describes the use of phenytoin as a treatment for PHM.

Lance Adam's Syndrome:

- Post-hypoxic Myoclonus
  - Defined as sudden muscular jerking movements of voluntary muscles
  - Initially, postural disorientation before treatment
  - Associated with diffuse neuronal degeneration, electrolyte imbalance
  - Improved rehabilitation techniques in increasing frequency.

Pathophysiology

- Myoclonus Jerks
  - Myoclonus: 
    - Hypersynchrony of somatosensory cortex or subcortical formation or both
    - Myoclonic: rapid, jerky, repetitive movements of one or more muscles
    - Myoclonus: intermittent, non-rhythmic
  - Myoclonus in Lance Adam's Syndrome: increased incidence of PHM

Diagnostic testing

- No radiologic study is specific
- Electroencephalography: myoclonic discharges, electrical silence
- Negative Myoclonus: interruption in muscle movement.
- Positive Myoclonus: myoclonic jerks, irregular or rhythmic

Therapeutic interventions

- Mediations are treatment of choice.
- Phenytoin: antiepileptic drug
  - Effective treatment for PHM
  - Decreases myoclonic jerks

Phenytoin

- Phenytoin was effective in this case.
- Reduced electrical conductance spanning brain areas.
- Stabilizes inactive state of sodium channels.
- Decreases transmembrane potential and increases potassium conductance.
- Reduces electrical conductance in neurons.

Phenytoin effects:

- Reduces electrical conductance spanning brain areas.
- Stabilizes inactive state of sodium channels.
- Decreases transmembrane potential and increases potassium conductance.
- Reduces electrical conductance in neurons.

Studies suggest trials of valproate, lamotrigine, levetiracetam for example.

Summary

- No specific intervention has been documented as effective.
- Confirmed diagnosis with careful history.
- Detailed mental status examination.
- Considerations include abnormal movements.
- Consider use of phenytoin if there is no response.
- Rehabilitation, with effective occupational therapy, can lead to improved function and quality of life for those struggling with PHM.

Post- hypoxic myoclonus (PHM) or Lance-Adams Syndrome, is a rare and difficult to treat condition that is characterized by involuntary muscular jerking movements in people who have sustained a cerebral event. Both the incidence and pathogenesis have not clearly been established, and little is known about effective treatments for this condition. This is a rare syndrome, and the successful treatment for PHM has been limited. This report is unusual in that it describes the use of phenytoin as a treatment for PHM.
Prevention of Limb Amputation as a Result of a Multidisciplinary Treatment Approach

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Introduction: A multidisciplinary treatment approach to the care of extensive lower extremity vascular wounds has prevented amputation in the long term acute care hospital (LTACH) setting. There is limited documentation in the literature as it relates to this clinical issue in LTACH's.

Clinical Problem: A patient with an extensive medical history including 25 pack years of smoking, peripheral vascular disease and painful claudication who underwent bifemoral bypass presented to the LTACH for continuing medical care. The patient's recovery in the acute care hospital was complicated by a thrombus for which a thrombectomy was done but there were already signs of necrosis. Further postop medical complications of renal failure, anemia, multiple wound infections and other skin breakdown occurred prior to admission to the long term acute care hospital. The patient was admitted to the LTACH with several extensive lower extremity vascular wounds including full thickness necrosis and exposure of bone and tendon on the toes, heels and distal lower extremities. The patient had considerable lower extremity edema that impeded healing. Initial dressing changes were painful and anxiety and depression were barriers to patient care.

Clinical Approach: A multidisciplinary management process consisted of chemical debridement of necrotic tissue, surgical debridement, hyperbaric oxygen therapy, pulsatile lavage by physical therapy, pain management, nutritional counseling for oral and parenteral nutrition, and psychology interventions for counseling and support of the patient and spouse. Treatment also included pressure relief and frequent repositioning on specialized support surface. When the wound beds were beginning to granulate, a Negative Pressure Wound Treatment was applied. The wound care team evaluated the patient weekly to monitor progress, modify the treatment plan as needed and to educate staff and patient.

Patient Outcomes: All but one heel wound completely closed, avoiding the need for bilateral above knee amputation. The patient has returned home, is ambulating and continues to have follow-up in an outpatient wound care center.

Conclusion: Superior outcomes were achieved due to the multidisciplinary treatment approach and advanced wound care modalities.
Zoledronic Acid in the Treatment of Immobilization Hypercalcemia in Spinal Cord Injury (SCI)

Case Report

A 28 year old gentleman sustained thoracic spine fractures and T5 ASIA B paraplegia from a motorcycle accident. He had multiple medical complications such as renal failure, dehydration, and pneumonia which delayed his return to rehabilitation. High calcium levels were noted 4 weeks after his injury, he was noted to have significant hypercalcemia. His work up was consistent with hypercalcemia from immobility, and included a low PTH, low 1,25 Vit D, high phosphate, elevated serum or phosphorus, normal PTH related peptide and a 24 hour urine calcium of 600 mg. There was no response to reduced calcium intake, rehydration with intravenous fluids, nor rehabilitation interventions for remobilization which included, for example, use of a standing frame, range of motion, transfer training and a full rehabilitation program, and eventual deterioration of the lower extremities. He received 4 mg of zoledronic acid intravenously. His calcium and phosphate normalized, and he remained ambulatory. The pathophysiology of hypercalcemia in SCI, the differential diagnosis, pathophysiology and pharmacologic treatment options are presented. The potential benefits and risks of zoledronic acid in comparison to other available agents are highlighted.

Hypercalcemia and Spinal Cord Injury

- Decreased muscle activity, decreased mechanical stimulation
- Most常见 in weeks, range 2 weeks to 6 months
- 10-25% frequency
- Common in older age vs paraplegia
- Increased urinary calcium excretion
- Less PTH leads to increased serum phosphate and decreased 1,25 Vit D
- More potent than pamidronate. It is the newest bisphosphonate, approved for the treatment of malignancy associated hypercalcemia, is a potent bisphosphonate that inhibits bone resorption. It is the newest bisphosphonate, and has been shown to be more effective than pamidronate in lowering calcium levels with a longer duration of response. However, there are no reports in the literature describing its use in immobilization hypercalcemia in SCI. The report is unusual in that it describes the use in hypercalcemia following spinal cord injury due to immobilization. A 28 year old gentleman sustained thoracic spine fractures and T5 ASIA B paraplegia from a motorcycle accident. He had multiple medical complications such as renal failure, pneumonia and required surgery which delayed his return to rehabilitation. High calcium levels were noted 4 weeks after his injury, he was noted to have significant hypercalcemia from immobility, and included a low PTH, low 1,25 Vit D, high phosphate, elevated serum or phosphorus, normal PTH related peptide and a 24 hour urine calcium of 600 mg. There was no response to reduced calcium intake, rehydration with intravenous fluids, nor rehabilitation interventions for remobilization which included, for example, use of a standing frame, range of motion, transfer training and a full rehabilitation program, and eventual deterioration of the lower extremities. He received 4 mg of zoledronic acid intravenously. His calcium and phosphate normalized, and he remained ambulatory. The pathophysiology of hypercalcemia in SCI, the differential diagnosis, pathophysiology and pharmacologic treatment options are presented. The potential benefits and risks of zoledronic acid in comparison to other available agents are highlighted.

Work up

- Serum Calcium greater than 14 mg/dl (3.5 mmol/L) requires treatment regardless of symptoms
- PTH may be normal in hyperparathyroid hypercalcemia
- Furosemide may be added to saline diuresis
- 12-14 mg/DL (3-3.5 mmol/L) may be well tolerated chronically
- Corrected serum calcium 24 hour urinary calcium excretion
- n-telopeptide or c-telopeptide biochemical markers of bone metabolism and most sensitive specific indicators of bone resorption
- Ionized calcium- best indicator
- • Diuresis
- • Calcium intake, rehydration with intravenous fluids, nor rehabilitation interventions for remobilization which included, for example, use of a standing frame, range of motion, transfer training and a full rehabilitation program, and eventual deterioration of the lower extremities. He received 4 mg of zoledronic acid intravenously. His calcium and phosphate normalized, and he remained ambulatory. The pathophysiology of hypercalcemia in SCI, the differential diagnosis, pathophysiology and pharmacologic treatment options are presented. The potential benefits and risks of zoledronic acid in comparison to other available agents are highlighted.

Zoledronic Acid

- Renest zoledronic acid approved for hypercalcemia of malignancy.
- More potent than pamidronate.
- More effective than pamidronate in lowering calcium levels with a longer duration of response. However, there are no reports in the literature describing its use in immobilization hypercalcemia in SCI. The report is unusual in that it describes the use in hypercalcemia following spinal cord injury due to immobilization. A 28 year old gentleman sustained thoracic spine fractures and T5 ASIA B paraplegia from a motorcycle accident. He had multiple medical complications such as renal failure, pneumonia and required surgery which delayed his return to rehabilitation. High calcium levels were noted 4 weeks after his injury, he was noted to have significant hypercalcemia from immobility, and included a low PTH, low 1,25 Vit D, high phosphate, elevated serum or phosphorus, normal PTH related peptide and a 24 hour urine calcium of 600 mg. There was no response to reduced calcium intake, rehydration with intravenous fluids, nor rehabilitation interventions for remobilization which included, for example, use of a standing frame, range of motion, transfer training and a full rehabilitation program, and eventual deterioration of the lower extremities. He received 4 mg of zoledronic acid intravenously. His calcium and phosphate normalized, and he remained ambulatory. The pathophysiology of hypercalcemia in SCI, the differential diagnosis, pathophysiology and pharmacologic treatment options are presented. The potential benefits and risks of zoledronic acid in comparison to other available agents are highlighted.

Conclusion

- • Work up is designed to assess degree of bone resorption
- • Treatment options include hydration and bisphosphonate use
- • Work up is designed to assess degree of bone resorption
- • Treatment options include hydration and bisphosphonate use
- • Less PTH leads to increased serum phosphate and decreased 1,25 Vit D
- • Bone turnover rate indicator
- • Corrected serum calcium 24 hour urinary calcium excretion
- • PTH levels- treated to low
- • 12-14 mg/DL (3-3.5 mmol/L) may be well tolerated chronically
- • Corrected serum calcium 24 hour urinary calcium excretion
- • n-telopeptide or c-telopeptide biochemical markers of bone metabolism and most sensitive specific indicators of bone resorption
- • Ionized calcium- best indicator
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Rhomboid/Levator Scapulae Spasticity After Incomplete Cervical Spinal Cord Injury Treated With Botulinum Toxin Type A: A Case Report

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**Patient**

This is a 76 year old male with C3 ASIA C spinal cord injury with restricted shoulder range of motion.

**Case Description**

The patient presented to the inpatient rehabilitation service two months after a fall with resultant central cord syndrome. Primary complaint was restriction in ability to abduct the shoulders. Physical examination revealed the medial borders of the scapulae were almost touching when the upper arms were resting against the chest wall. Rhomboids and levator scapulae were found to be strongly contracting with any active or passive movement of the shoulder, which led to further adduction of the scapulae. Left scapula had no winging and 3/5 serratus anterior strength. Right scapulae had significant medial winging, and 1/5 serratus anterior strength. Shoulder abduction was restricted to 60 degrees bilaterally. Aggressive stretching, heat, taping, and electrical stimulation were unsuccessful in improving range of motion over a six week period during an inpatient rehabilitation stay. Botulinum toxin Type A was injected into the bilateral rhomboids and levator scapulae, and reassessed at 2 weeks and 3 and 6 months post-injection.

**Conclusions**

In this case, the left side showed improvement due to the ability of the serratus anterior to rotate the glenoid upwards and allow further humeral abduction. The lack of serratus anterior strength on the right side led to no improvement in range of motion. The one time use of Botulinum Toxin Type A led to a reduction in spasticity, improvement in scapular position at rest, range of motion of the shoulder, as well as functional improvements. Evaluation of the posterior shoulder musculature in the setting of reduced active and passive shoulder range of motion can lead to the identification of treatable causes and improve patient outcomes.

**KEYWORDS:** Spinal Cord Injuries, Rehabilitation, Muscle Spasticity, Botulinum Toxin Type A.
**Negative Pressure Wound Therapy as an Alternative to Surgical Flap Closure**

**Introduction:** The etiology of wounds that are treated in the Long Term Acute Care Hospital (LTACH) varies greatly. These patients have multiple medical and surgical issues that in many cases result in difficult to close pressure ulcers, surgical wounds and those wounds as a result of acute on chronic illnesses. The treatment of these wounds remains a challenge.

**Clinical Problem:** For some wounds, one of the most effective and timely means of closure is by undergoing plastic surgery. When flap closure is an option from a clinical perspective it may not be an option from a resource perspective. We need to be prepared to utilize alternate treatment such as Negative Pressure Wound Therapy (NPWT). NPWT in conjunction with other wound treatments has been shown to provide outstanding outcomes in patients in the LTACH setting (Trigilia et al. SAWC/WOCN, 2009).

**Clinical Approach:** Upon admission the Wound Care team evaluation resulted in the development of a treatment plan that included a request for a plastic surgery consult. The plastic surgeon felt that 2 out of the 3 patients could benefit from surgery, but other circumstances could not support this treatment plan. At that time the team initiated an alternative treatment plan with NPWT.

**Patient Outcomes:**

**Patient # 1: Male with Spinal cord Injury.**
A.P. is a 42 year old man who sustained several gunshot wounds including one in the left buttock and trauma resulting in a spinal cord injury. He had a large wound when he was admitted to Gaylord. Plastic surgery was consulted. Debridement was performed and NPWT initiated.

NPWT for 12 weeks and healing progresses from 7.5cm x 6.5cm x 2.5cm with undermining of 2.0cm to 0.7cm x 1.5cm x 1.0cm from 9/03/09 to 12/03/09.

**Patient # 2: Female with scoliosis repair, infection, respiratory failure and diabetes.**

Patient underwent extensive thoracic spine surgery in 11/2008 for severe scoliosis. In 02/02/09, she developed back pain and L.1 scan showed multiple paraspinal abscesses along the length of her thoracic spine.

B.R. underwent removal and replacement of the Harrington rods and a large wound remained to close by secondary intention. She was admitted to Gaylord LTACH on 5/20/09. Surgical intervention to close the wound was not possible and hardware visible to present measurements; proximal opening 13.7cm x 4.2 cm x 3.0 cm and distal opening 12.6 cm x 2.5cm x 0.1 cm with a large area of closure in between. After almost 10 months of NPWT with partial closing of the wound, flap surgery was done.

**Patient # 3: Male with spinal cord injury with failed gluteal flap.**
R. S. is a 51 year old male with tetraplegia who underwent an excision and rotational flap surgery for left ischial tuberosity stage IV pressure ulcer on 9/1/09. He returned to Gaylord on 09/03/09, with partial dehiscence left IT incision. Patient seen on admission after the flap and there was some separation of the sutures distally. Patient was started on pulse lavage to maintain a clean wound bed and remove debris. NPWT was also started at this time. Approximately 35 weeks of NPWT the wound healing progressed from 44.5cm x 6.5 cm x 2.5cm to 3.0 cm x 4.0cm x 0.1 cm with deminishing of 2.0cm from 06/19/09 to 04/20/10.

NPWT for 11 weeks with healing progresses from 6.2cm x 2.0cm x 1.5cm to 0.7cm x 1.5cm x 1.0cm.

**Conclusion:** In the LTACH setting is has been shown that NPWT is advanced wound treatment that has demonstrated positive outcomes for those patients not appropriate for surgical closure of their wounds.