

# An Unnerving Situation

Joel Burnett, MD

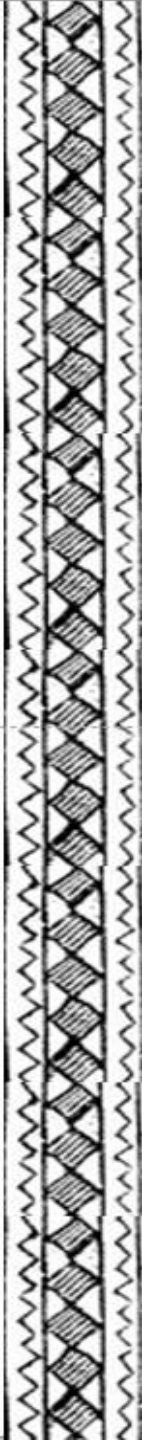


*Alice's Adventures in Wonderland*  
by Lewis Carroll

Illustrations by Sir John Tenniel



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61-year old woman with history of:

- CKDIII
- Inflammatory breast cancer (2007) in remission
- Herpes genitalis
- Chronic lumbar back pain
- Tobacco use disorder



One last thing, doc.  
I was too embarrassed to say this before...



...I'm going to the bathroom when I don't mean to.



...I'm going to the bathroom when I don't mean to.  
**Bowel and bladder incontinence**



...I'm going to the bathroom when I don't mean to.

Bowel and bladder incontinence

Perineal numbness





...I'm going to the bathroom when I don't mean to.

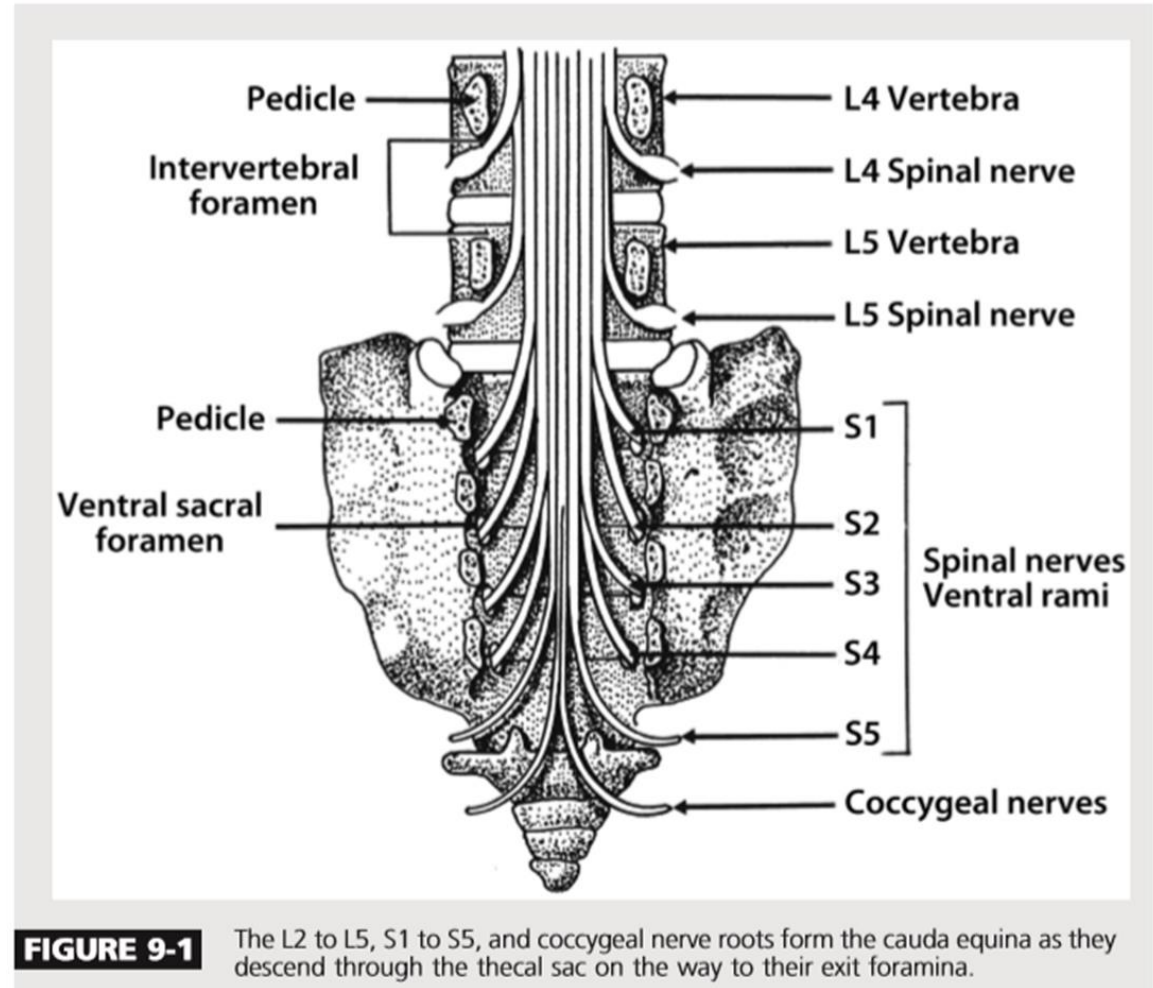
Bowel and bladder incontinence

Perineal numbness

Back pain



# Cauda Equina Syndrome



# Cauda Equina Syndrome: A Literature Review of Its Definition and Clinical Presentation

*Stuart Fraser, BSc, Lisa Roberts, PhD, Eve Murphy, MSc*

## Defining Cauda Equina Syndrome

For a diagnosis of CES, one or more of the following must be present: (1) bladder and/or bowel dysfunction, (2) reduced sensation in the saddle area, or (3) sexual dysfunction, with possible neurologic deficit in the lower limb (motor/sensory loss, reflex change).



# Differential Diagnosis

## Compressive

- Disk herniation
- Fracture
- Neoplasms
- Iatrogenic injury
- Chronic lumbosacral spinal stenosis
- Infection



# Differential Diagnosis

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- Disk herniation
- Fracture
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- Infection

## Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection



# Focused Neurologic Exam

- Motor Strength: 5/5 upper and lower extremities
- Reflexes: 2+ upper and lower extremities
- Pathologic Reflexes: absent Babinski sign and clonus
- Sensation: **perianal sensation diminished on left**
- Anal Sphincter: **flaccid rectal tone**
- Gait: normal



# Problem-based History

- Chronic lumbar **back pain** with left-sided radiculopathy that was **unchanged from baseline**
- New-onset **acute** perineal numbness, urinary retention and constipation, then days later **urinary and fecal incontinence**, now only fecal incontinence
- 6-7 week duration of symptoms, **improving**
- No identifiable inciting event, trauma, fever, night sweats, weight loss, or IV drug use

# Laboratory Data

Na: 139 mmol/L    Hgb: 11.4 g/dL  
K: 4.4 mmol/L    WBC: 5.5 K/cu mm  
Cl: 100 mmol/L    Plt: 202 K/cu mm  
CO2: 27 mmol/L  
BUN: 21 mg/dL    Ca 27-29: 23.8 U/mL  
Cr: 1.4 mg/dL  
Ca: 9.5 mg/dL  
AST: 28 U/L  
ALT: 18 U/L  
Alk Phos: 40 U/L  
Total Bilirubin: 0.6 mg/dL  
Total Protein: 7.1 g/dL  
Albumin: 3.7 g/dL

# Imaging

CXR: clear lungs  
Mammogram: unremarkable





# Problem Representation #1

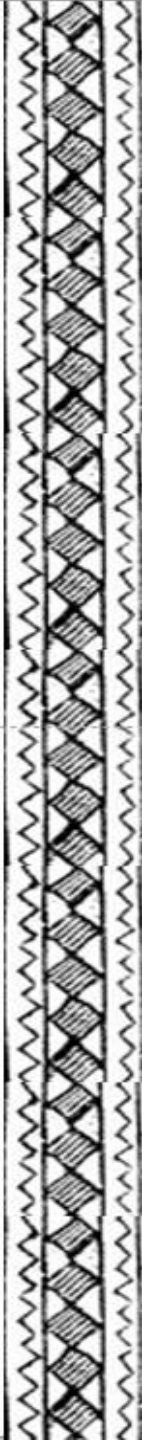
61-year-old woman with history of tobacco use disorder, chronic lumbar back pain with radiculopathy, and breast cancer in remission presenting with cauda equina syndrome.



# Prioritized Differential Diagnosis

- Chronic, progressive spinal stenosis
- Recurrent breast cancer with metastasis
- Disk herniation
- New malignancy





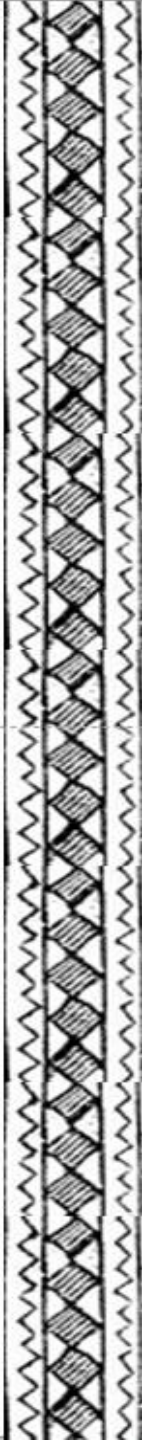
“Her degree of stenosis does not explain her incontinence symptoms. Other causes should be investigated.”

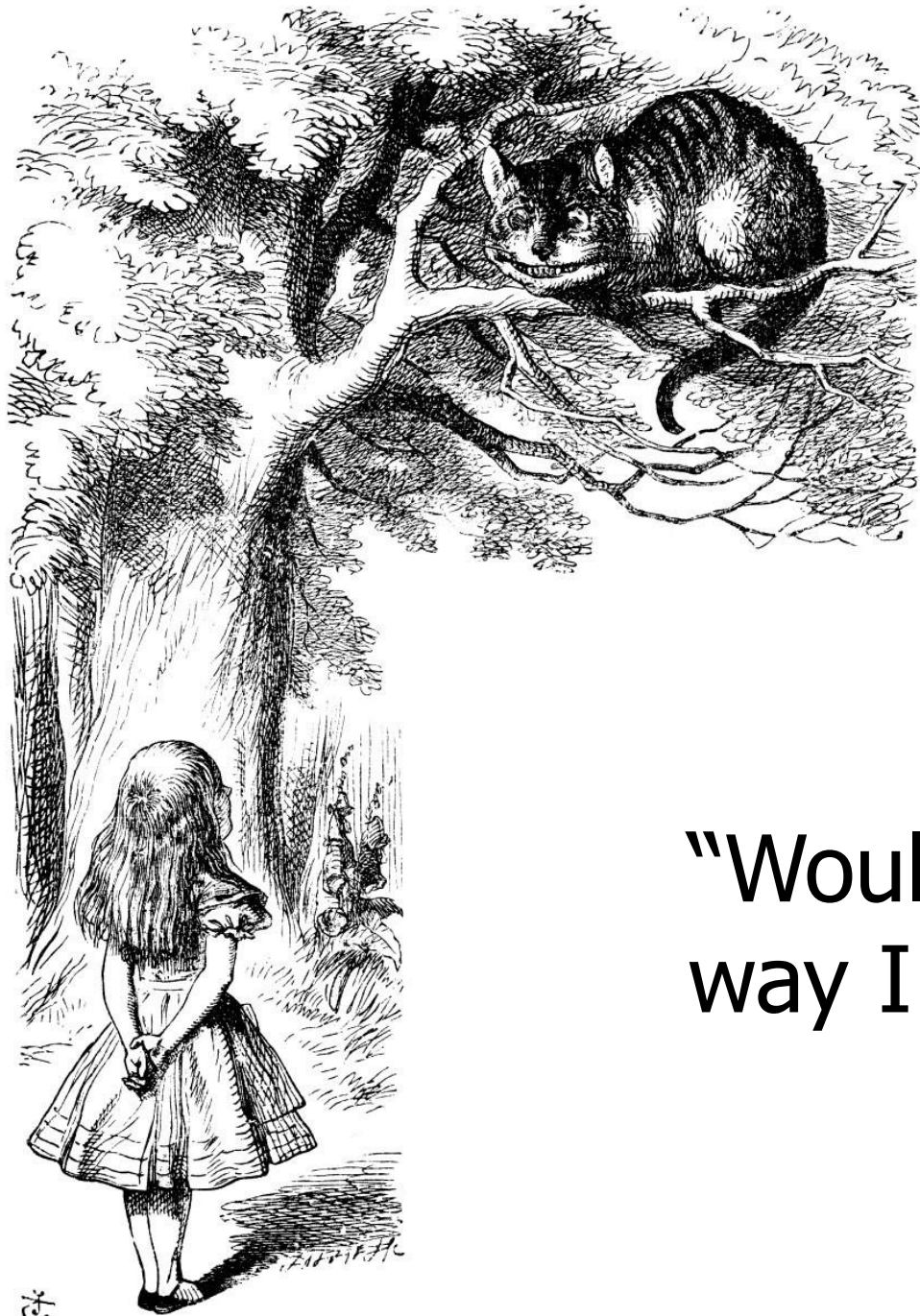




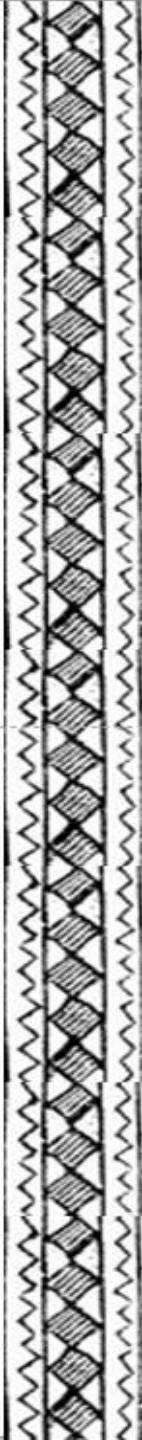
2/4

W. P. LITTLE





“Would you tell me, please, which way I ought to go from here?”



## Problem Representation #2

61-year-old woman presenting with **acute onset** of cauda equina syndrome with **progressive, spontaneous** symptom **improvement** over 6 weeks.



# Differential Diagnosis

## Compressive

- Disk herniation
- Fracture
- Neoplasms
- Iatrogenic injury
- Chronic lumbosacral spinal stenosis
- Infection

## Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection





# Differential Diagnosis

## Compressive

- Disk herniation
- Fracture
- Tumors
- Traumatic injury
- Chronic lumbosacral spinal stenosis
- Infection

## Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection



# Key Features

- Acute onset
- Self-limited
- Non-compressive



GH

# Differential Diagnosis

## Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection



# Differential Diagnosis

## Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection

- Dural AV Fistula
- Vascular Accident



# Differential Diagnosis

## Non-compressive

- Ischemia
  - Infiltration
  - Inflammation
  - Infection
- Sarcoidosis



# Differential Diagnosis

## Non-compressive

- Ischemia
  - Infiltration
  - Inflammation
  - Infection
- Guillain-Barré
  - Vasculitis



# Differential Diagnosis

## Non-compressive

- Ischemia
  - Infiltration
  - Inflammation
  - Infection
- Neurosyphilis
  - Tuberculosis
  - CMV + AIDS
  - VZV
  - HSV



**HSV**





61-year old woman with history of:

- CKDIII
- Inflammatory breast cancer (2007) in remission
- Herpes genitalis
- Chronic lumbar back pain
- Tobacco use disorder



# Outside Records – Urgent Care Visit

- Chief complaint: vaginal and pelvic pain
- HPI: onset 1 week prior to cauda equina syndrome
- GU Exam: multiple, bilateral ulcerations of the labia and surrounding perineum
- Vaginal Swab: + Herpes Simplex Virus

## Problem Representation #3

61-year-old woman presenting with **acute**, primary **genital herpes infection**, followed days later by **acute** onset of **cauda equina syndrome**, with **progressive symptom improvement** over 6 weeks.



# Elsberg Syndrome

An infectious syndrome consisting of acute or subacute lumbosacral radiculitis, with or without myelitis, provoked by primary or reactivated herpes simplex infection



# Elsberg Syndrome – Diagnostic Criteria

A. Required
A1. Clinical symptoms and signs of cauda equina involvement: urinary hesitancy or retention; bowel incontinence, or severe constipation (erectile dysfunction insufficient on its own) (n = 39, 80%)
A2. MRI or electrophysiologic evidence of cauda equina involvement: enhancement of cauda equina; EMG evidence of radiculopathy (n = 20, 41%)
B. Supportive but not required
B1. Time course: acute/subacute onset; no relapse; progression over less than 3 mo (n = 42, 86%)
B2. Coexisting or recently preceding symptoms of genital herpes infection OR other clinical symptoms of herpes virus infection (n = 5, 10%)
B3. Clinical (e.g., exaggerated reflexes and Babinski signs) or MRI evidence of myelitis in conus (n = 28, 57%)
B4. CSF pleocytosis (n = 23, 47%)
B5. Documented herpes virus infection from CSF by PCR, culture, or detection of IgM serology (n = 5, 10%)
C. Red flags
C1. Relapses beyond 1 y from onset (n = 4, 8%)
D. Exclusionary
D1. Myelitis extending rostral to T9 (n = 12, 25%)
D2. Other neurologic symptoms suggestive of alternative etiology: optic neuritis, brain/brainstem syndrome (n = 3; 6%)
D3. Other etiology proven/more likely for syndrome: NMOSD, dural arteriovenous fistula, viral transverse myelitis, other causes of myelopathy (n = 11, 22%)

- Clinical signs/symptoms of cauda equina syndrome

# Elsberg Syndrome – Diagnostic Criteria

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- Acute/subacute onset; no relapse
- Coexisting or recently preceding HSV infection

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- MRI evidence of cauda equina involvement

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- Clinical or MRI evidence of myelitis
- CSF pelocytosis
- HSV infection from CSF by PCR, culture, or IgM



# Elsberg Syndrome – Natural History

- Neurologic clinical spectrum of HSV genital infections:
  - urinary retention (seen in 5-15% of primary infections)
  - aseptic meningitis
  - cauda equina syndrome
  - myelitis
- Self-limited, but may leave some degree of permanent neurologic impairment
- Treatment: in acute period, IV acyclovir +/- steroids

# Follow Up

- History: continued symptom improvement
- Labs: negative HIV and syphilis
- Plan:
  - Offered consultation with neurology and additional diagnostics – she declined
  - Monitored for new neurologic symptoms – none to date
  - Suppressive acyclovir



“Tut, tut, child!” said the Duchess. “Everything’s got a moral, if only you can find it.”



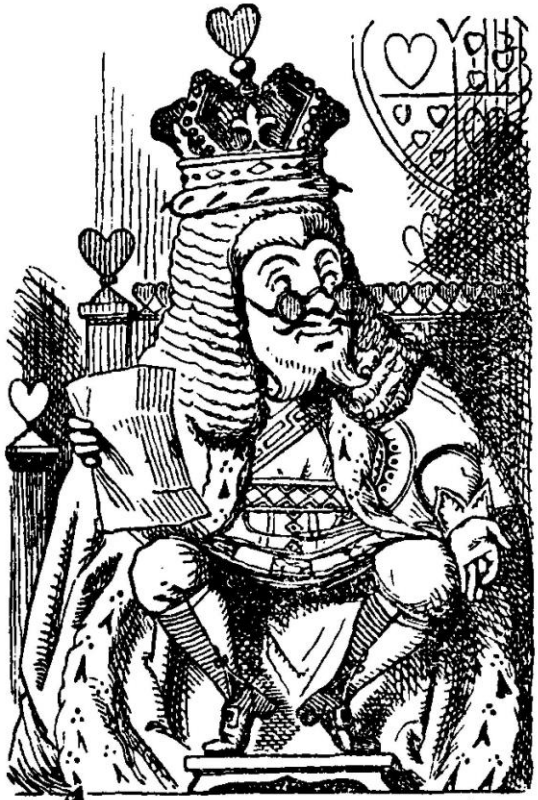
# Moral of the Story

- Not all causes of cauda equina syndrome are surgical emergencies (though we should proceed as if they are)
  - non-compressive etiologies include ischemia, infection, inflammation, and infiltration

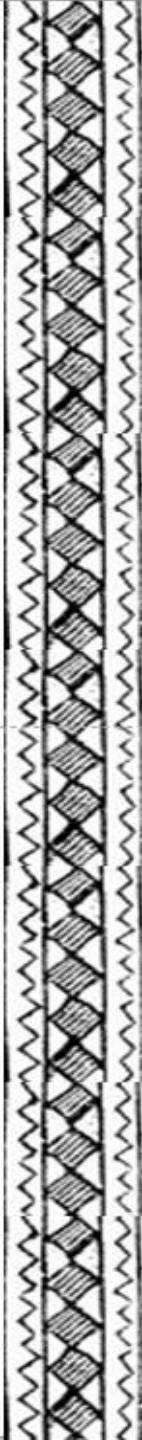
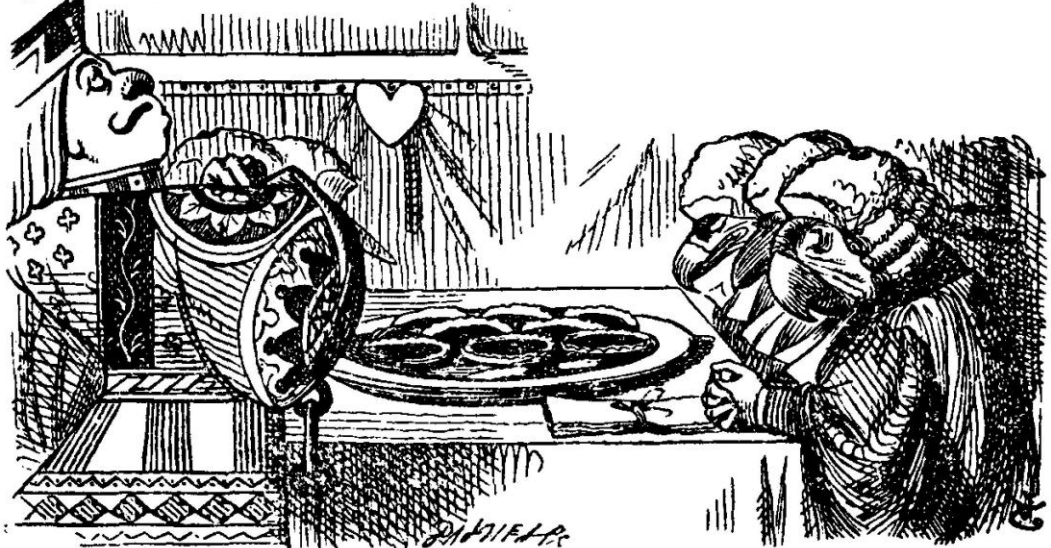


# Moral of the Story

- Not all causes of cauda equina syndrome are surgical emergencies (though we should proceed as if they are)
  - non-compressive etiologies include ischemia, infection, inflammation, and infiltration
- Elsberg Syndrome is an uncommon manifestation of a common infection
  - Clinical spectrum of HSV genital infection includes radiculitis and myelitis
  - Diagnosis may be aided by laboratory evidence of HSV infection, CSF studies, and MRI



“Begin at the beginning,” the king said, very gravely, “and go on till you come to the end: then stop.”





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# Elsberg Syndrome - Epidemiology

- Most commonly caused by HSV-2
- Most often seen in sexually active young women
- Under-recognized; data limited to case reports and series



