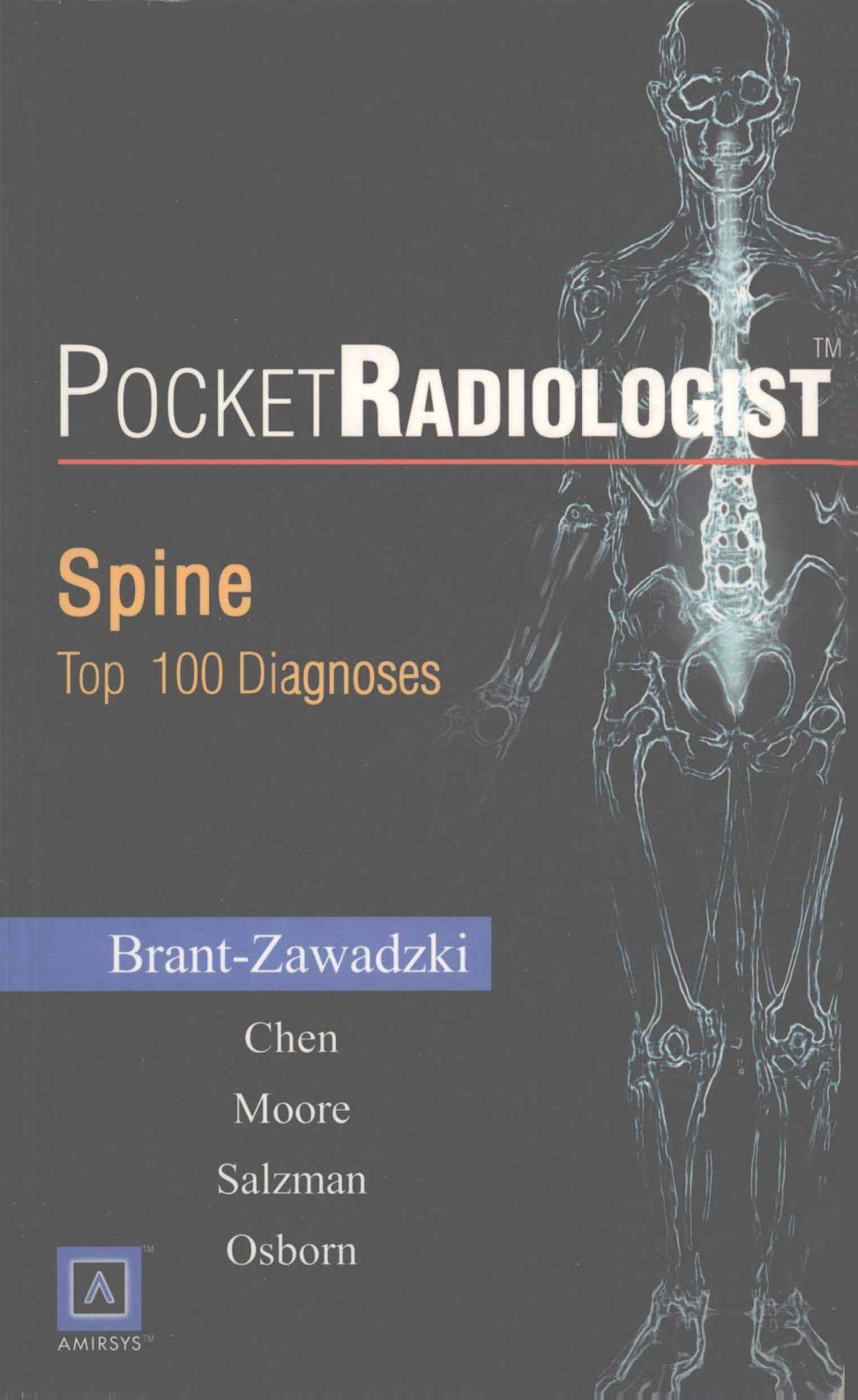

POCKET RADIOLOGIST™

Spine

Top 100 Diagnoses

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Spine
100 Top Diagnoses

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Preface

The **PocketRadiologist™** series is an innovative, quick reference designed to deliver succinct, up-to-date information to practicing professionals "at the point of service." As close as your pocket, each title in the series is written by world-renowned authors, specialists in their area. These experts have designated the "top 100" diagnoses in every major body area, bulleted the most essential facts, and offered high-resolution imaging to illustrate each topic. Selected references are included for further review. Full color anatomic-pathologic computer graphics model many of the actual diseases.

Each **PocketRadiologist™** title follows an identical format. The same information is in the same place—every time—and takes you quickly from key facts to imaging findings, differential diagnosis, pathology, pathophysiology, and relevant clinical information.

PocketRadiologist™ titles are available in both print and hand-held PDA formats. Our first modules feature Spine, Head and Neck, and Orthopedic (Musculoskeletal) Imaging. Additional titles include Spine and Cord, Chest, Breast, Vascular, Cardiac, Pediatrics, Emergency, and Genital Urinary, and Gastro Intestinal. Enjoy!

Anne G Osborn MD
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PocketRadiologist™
Spine
Top 100 Diagnoses

The diagnoses in this book are divided into 11 sections in the following order:

Congenital
Trauma
Degenerative
Infections
Inflammatory/Autoimmune
Neoplasms
Non-Neoplastic Cysts and Masses
Post Operative Complications
Vascular Lesions
Vertebral Marrow Changes
Peripheral Nerve/Plexus Imaging

Table of Diagnoses

Congenital

Trauma

Degenerative

Infections

Inflammatory / Autoimmune

Neoplasms

Non-Neoplastic Cysts and Masses

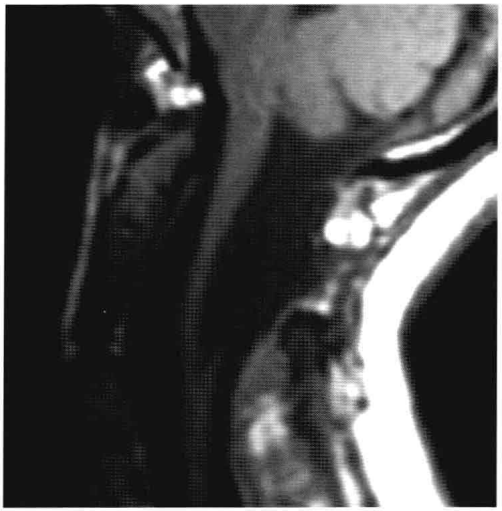
Post Operative Complications

Vascular Lesions

Vertebral Marrow Changes

Peripheral Nerve / Plexus Imaging

Neurenteric Cyst



Sagittal T1WI of the cervical spine demonstrates fused upper cervical vertebral bodies. There is a capacious fluid-filled space in the posterior upper cervical canal.

Key Facts

- Synonym: Enterogenous cyst
- Definition: Intraspinal enteric-lined cyst
- Classic imaging appearance: Intradural extramedullary cyst with associated vertebral anomalies
- Other key facts
  - Along the spectrum of split notochord syndrome
  - Most common in the anterior thoracic (42% in one series) or cervical spine (32%), rare in lumbar spine
  - Usually midline
  - Vertebral anomalies including spina bifida, vertebral fusion, butterfly vertebra, or scoliosis in about half of the cases

Imaging Findings

General Features

- Best imaging clue: Vertebral anomalies with an intraspinal cyst

CT Myelogram Findings

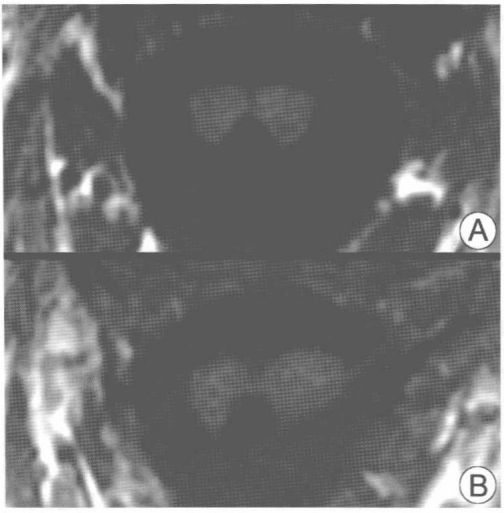
- Vertebral anomalies
- Focal enlargement of the canal
- Intradural extramedullary cyst
- Invaginating cyst may mimic an intramedullary lesion

MR Findings

- Well-circumscribed, intradural, extramedullary fluid-intensity lesion
  - Iso- to hyperintense to cerebral spinal fluid (CSF) on T1WI and T2WI depending on protein content
  - No enhancement after contrast injection
  - Focal cord atrophy from chronic mass effect

Imaging Recommendations

- Coronal T1WI to better assess vertebral anomalies



Axial T1WI (A, B) of the upper cervical spine reveals a cyst isointense to CSF, splaying the posterior cervical cord at the midline.

Differential Diagnosis

Arachnoid (Meningeal) Cyst

- CSF intensity on all pulse sequences
- Primary arachnoid cyst located posteriorly in the spinal canal
- Secondary arachnoid cyst no specific preference
- Lacks vertebral anomalies

(Epi)dermoid Cyst

- Usually in the lumbar spine
- May see sinus tract (20%) or cord tethering

Pathology

General

- Embryology-Anatomy
  - During the third week of embryonic life, the notochord forms and separates the dorsal ectoderm (skin and spinal cord) and the ventral endoderm (foregut)
  - Failure of separation results in a split notochord or a notochord deviated to the left or right of the adhesion
- Etiology-Pathogenesis (spectrum of split notochord syndrome)
  - Dorsal enteric fistula
    - Most severe
    - Connecting the intestinal cavity with the dorsal skin surface, traversing through soft tissues and spine
    - Part(s) of the fistula may obliterate, forming other anomalies
  - Dorsal enteric sinus
    - Blind ending tract with opening on the dorsal skin surface
  - Dorsal enteric enterogenous cyst
    - Prevertebral, intraspinal, postvertebral, mediastinal, or mesenteric in location

- Dorsal enteric diverticulum
  - Diverticulum from the dorsal mesenteric border of the bowel
- A combination of the above anomalies may be present in one patient

- Epidemiology
  - Second to fourth decade of life
  - M: F = 3:2

Microscopic Features

- Thin-walled cyst lined by simple, pseudostratified, or stratified cuboidal or columnar epithelium
- Ciliated epithelium and goblet cells may be present
- Clear or proteinaceous fluid

Clinical Issues

Presentation

- Back pain
- Progressive paraparesis and paresthesia
- Gait disturbance

Natural History

- Progressive neurological deterioration

Treatment

- Surgical excision
- Drainage and partial resection if complete excision not possible

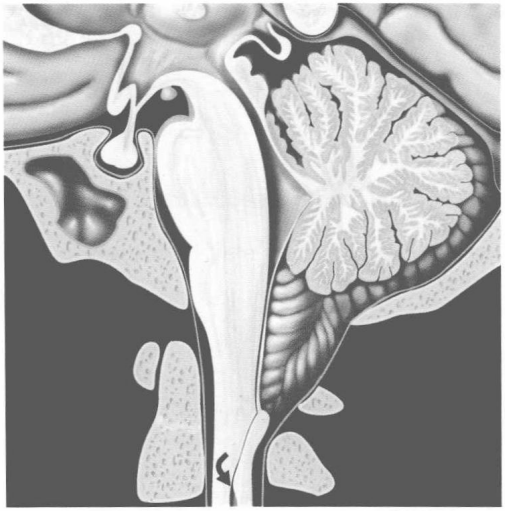
Prognosis

- Significant symptomatic improvement

Selected References

1. Barkovich AJ: Pediatric Neuroimaging. 2nd ed. 510-3, 1995
2. Gao PY et al: Neurenteric cysts: pathology, imaging spectrum, and differential diagnosis. International Journal of Neuroradiology 1:17-27, 1995
3. Geremia GK et al: MR imaging characteristics of a neurenteric cyst. AJNR 9:978-80, 1988

Chiari I, Spine



Sagittal midline graphic depicts Chiari I. Note "peg-like", low-lying tonsil with more vertically-oriented sulci. A collapsed syrinx is illustrated (curved arrow). The 4th ventricle is normal.

Key Facts

- Definition: Cerebellar tonsils extending below the foramen magnum
- Classic imaging appearance: Pointed cerebellar tonsils 5 mm below the foramen magnum, with associated syringohydromyelia
- Caused by mild "mismatch" between posterior fossa size (small), cerebellum (normal) ≥ tonsillar "ectopia"
- Tonsils can normally lie below foramen magnum (5 mm or less in adults, slightly more in children < 4y)
- Unless tonsils > 5 mm and/or pointed, probably not Chiari I

Imaging Findings

General Features

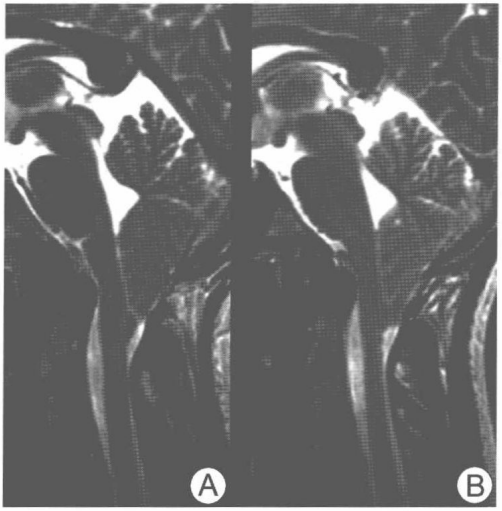
- Best imaging clue: Low-lying, pointed (not round), "peg-like" tonsils with vertical (not horizontal) sulci
- 4th occipital sclerotome syndromes (short clivus, craniovertebral segmentation/fusion anomalies) in 50%

CT Findings

- "Crowded" foramen magnum
- Small/absent PF cisterns
- Lateral/3rd ventricle is usually normal
  - +/- Ventriculomegaly
  - Depends upon degree of foramen magnum impaction

MR Findings

- Pointed, triangular-shaped ("peg-like") tonsils
  - ≥ 5 mm below foramen magnum **or**
  - Loss of normal round shape
  - Surrounding CSF effaced
- Small bony PF ≥ low torcular, effaced PF cisterns
- Short clivus ≥ apparent descent 4th ventricle, medulla
  - May be real if LP shunt present
- +/- Syringohydromyelia (14%-75%)



Sagittal T2WIs (A,B) in an asymptomatic patient show findings of classic Chiari I malformation. Note pointed, "peg-like" tonsils 10 mm below foramen magnum. The tonsillar sulci are oriented almost vertically.

Other Modality Findings
- Phase-contrast CSF flow/cord motion MR
  - Demonstrates pulsatile systolic tonsillar descent
  - Obstructed CSF flow across foramen magnum

Imaging Recommendations
- MR brain +/- CSF flow studies
- Image the spine to look for
  - Syrinx, low/tethered cord, or fatty filum

Differential Diagnosis

Acquired Tonsillar Ectopia/Herniation
- Basilar invagination
- "Pull from below": LP/LP shunt ⇒ intracranial hypotension with "sagging" brainstem, acquired tonsillar herniation
- "Push from above"
  - Chronic VP shunt
    - Look for thick skull, premature sutural fusion
    - Arachnoidal adhesions common
  - Tonsillar herniation 2° ↑ICP, mass effect

Pathology

General
- Genetics
  - Syndromic/familial
    - Velocardiofacial/microdeletion chromosome 22
    - Williams syndrome
    - Craniosynostosis
- Embryology
  - Underdeveloped occipital enchondrium ⇒ small posterior fossa vault ⇒ crowded PF ⇒ downward herniated hindbrain ⇒ obstructed

- Foramen magnum ⇒ lack of communication between cranial/spinal CSF compartments
- Etiology-Pathogenesis-Pathophysiology
  - Hydrodynamic theory of symptomatic Chiari I
    - Systolic piston-like descent of impacted tonsils/medulla ⇒
    - **Abnormal** pulsatile intraspinal CSF pressure-wave
    - May lead to hydrosyringomyelia
- Epidemiology = 0.01% of population

Gross Pathologic, Surgical Features

- Herniated, sclerotic tonsillar pegs; tonsils grooved by opisthion

Microscopic Features

- Purkinje/granular cell loss

Staging or Grading Criteria

- I = asymptomatic: ≈ 14-50%, treatment controversial
- II = brainstem compression
- III = hydrosyringomyelia

Clinical Issues

Presentation

- Up to 50% asymptomatic
- May mimic multiple sclerosis!
- "Chiari I spells": Cough/headache/ sneeze/syncope
- Symptomatic brainstem compression
  - Hypersomnolence/central apnea/(infant), sudden death
  - Bulbar signs (e.g., lower CN palsies)
  - Neck/back pain, torticollis, ataxia
- Symptomatic syringohydromyelia
  - Paroxysmal dystonia, unsteady gait, incontinence
  - Atypical scoliosis (progressive, painful, atypical curve)
  - Dissociated sensory loss/neuropathy (hand muscle wasting)

Natural History

- Increasing ectopia + ↑ time ⇒ ↑ likelihood symptoms
- Children respond better than adults; treat early

Treatment & Prognosis

- Controversial: Intervention for asymptomatic Chiari I + syrinx
- Direct shunting of symptomatic syrinx obsolete
- Aim = restore normal CSF flow at/around foramen magnum
  - PF decompression/ resection posterior arch C1
    - >90% ↓ brainstem signs
    - >80% ↓ hydrosyringomyelia
    - Scoliosis arrests (improves in youngest)
  - +/- Duraplasty, cerebellar tonsil resection
- Anterior decompression/posterior stabilization rarely indicated (some craniocervical anomalies)

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