THE CHIARI MALFORMATIONS: DIAGNOSIS AND MANAGEMENT

Historical overview

- Named after Hans Chiari (1851–1916) .
- Professor of Pathology in Prague, Czechoslovakia,
- Paper entitled “Concerning alterations in the cerebellum resulting from cerebral hydrocephalus”
- Published in Deutsche Medizinische Wochenscriff in 1891
- Autopsy series, 40 cases
- Described cerebellar anomalies in congenital hydrocephalus

- Initially described three malformations
- 5 years later he revised the second and added a fourth
- Arnold described a single case with Chiari II features
- The chiari II malformation also known as Arnold-Chiari malformation.

Introduction and definition

- Series of hindbrain anomalies
- No anatomical or embryological correlation between them
- Four types

Type	Definition
Chiari I	Caudal descent of *cerebellar tonsils* > 5mm below foramen magnum Hydrocephalus uncommon
Chiari II	Caudal herniation of *cerebellar vermis, brainstem and fourth ventricle* Almost all have hydrocephalus and myelomeningocele
Chiari III	Chiari II and posterior fossa contents herniating into occipital/ high cervical encephalocele
Chiari IV	Cerebellar aplasia or hypoplasia with aplasia of tentorium cerebelli

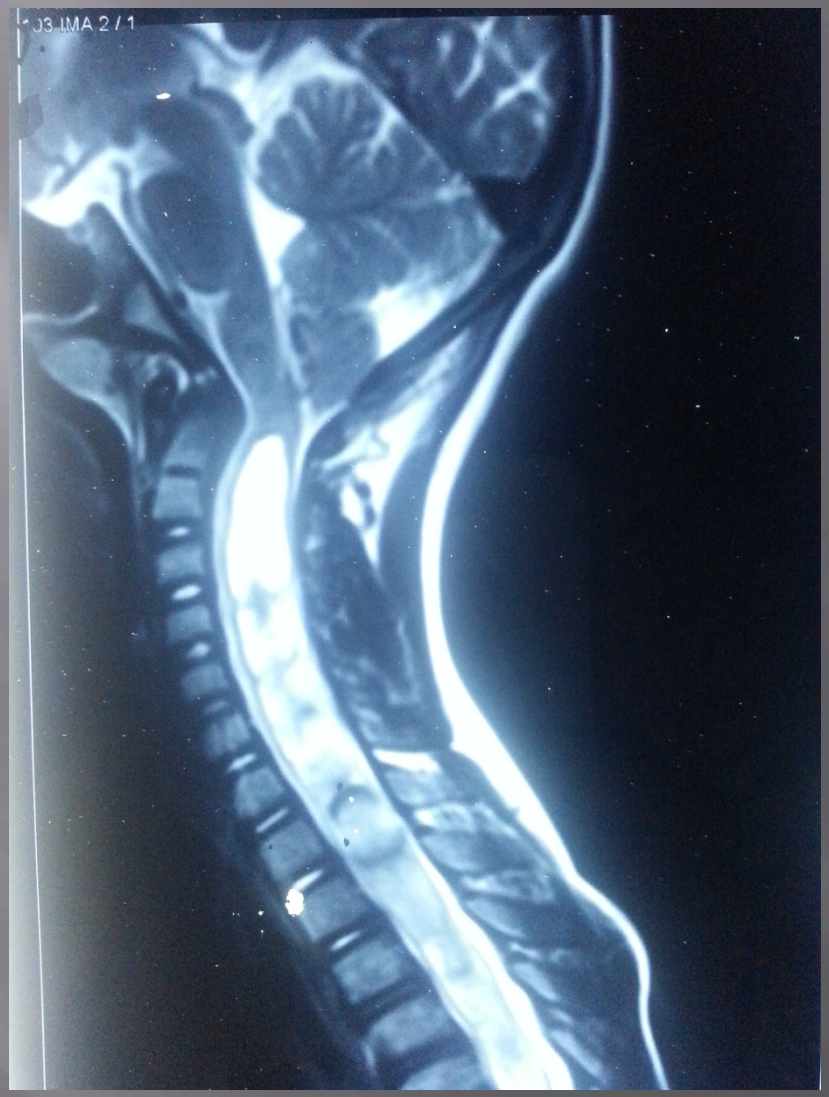
Chiari I malformation

- Traditionally defined as > 5mm tonsillar descent below the foramen magnum.
- Tonsils ascend with age
- Abnormal for age
  - > 6mm in first decade
  - > 5 mm in second and third decade
  - > 4 mm in fourth through eighth decade
  - > 3 mm in ninth decade.

- More important than absolute tonsillar descent may be
  - Peg like shape of tonsils
  - Attenuation of posterior fossa cisternal spaces
  - Suggestive clinical picture
- Primarily Chiari I is manifestation of underdevelopment and malformation of occipital cranium
- Cerebellar ectopia due to reduced posterior fossa volume and crowding of contents

- Other manifestations of hypodeveloped post fossa
  - Increased slope of the tentorium
  - Reduced height of the supraocciput
  - Reduced length of the clivus
  - Retroflexion of odontoid process

- Acquired tonsillar ectopia
  - Due to reduced volume of cranial cavity (Rickets, Craniosynostosis, Pagets disease)
  - Due to increase in volume of intracranial contents (acute hydrocephalus, tumour, cerebral edema)



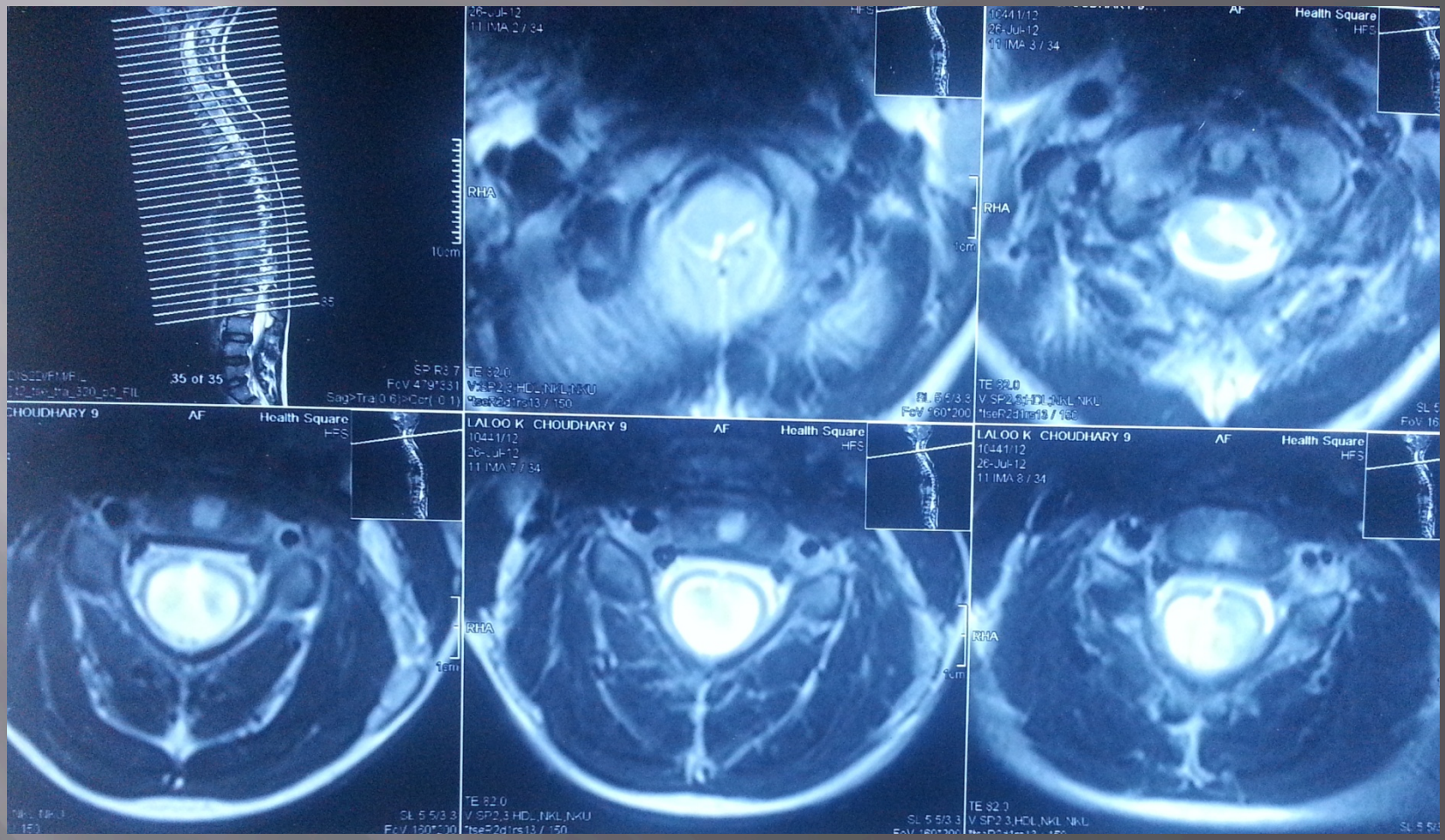
Chiari I malformation

Associated findings

- Skull
  - Shortened supraocciput
  - Shortened clivus
  - Larger than normal foramen magnum
  - Empty sella
  - Clival concavity, platybasia, basilar impression

- Spine
  - Klippel-flail deformity and atlanto axial assimilation
  - Retroflexed odontoid process
  - Thickened ligamentum flavum
  - Scoliosis
- Meninges
  - Elevated slope of tentorium cerebelli
  - Thickened arachnoid at foramen magnum level
  - Dural thickening/ at the level of arch of atlas

- Spinal cord
  - 50 to 75 % have cavitation within the cord (syrinx)
  - Lower cervical and thoracic cord mostly involved.
  - Segment of cord caudal to 4^{th} ventricle may be spared from cavitation.
- Brain usually normal except for tonsillar abnormality
- Hydrocephalus described in 3 to 10 %.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Signs and symptoms

- Pain is the most common complaint
  - Occipital and cervical region pain aggravated by Valsalva, cough-laugh headaches
- Signs and symptoms related to brainstem / cranial nerve and cerebellar compromise
  - Ataxia, downbeating nystagmus, incordination, dizziness
  - Dyspahgia, dysarthria, hiccoughs , glossal atrophy
  - Impaired gag, facial numbness
  - Extreme cases – “cerebellar fits”

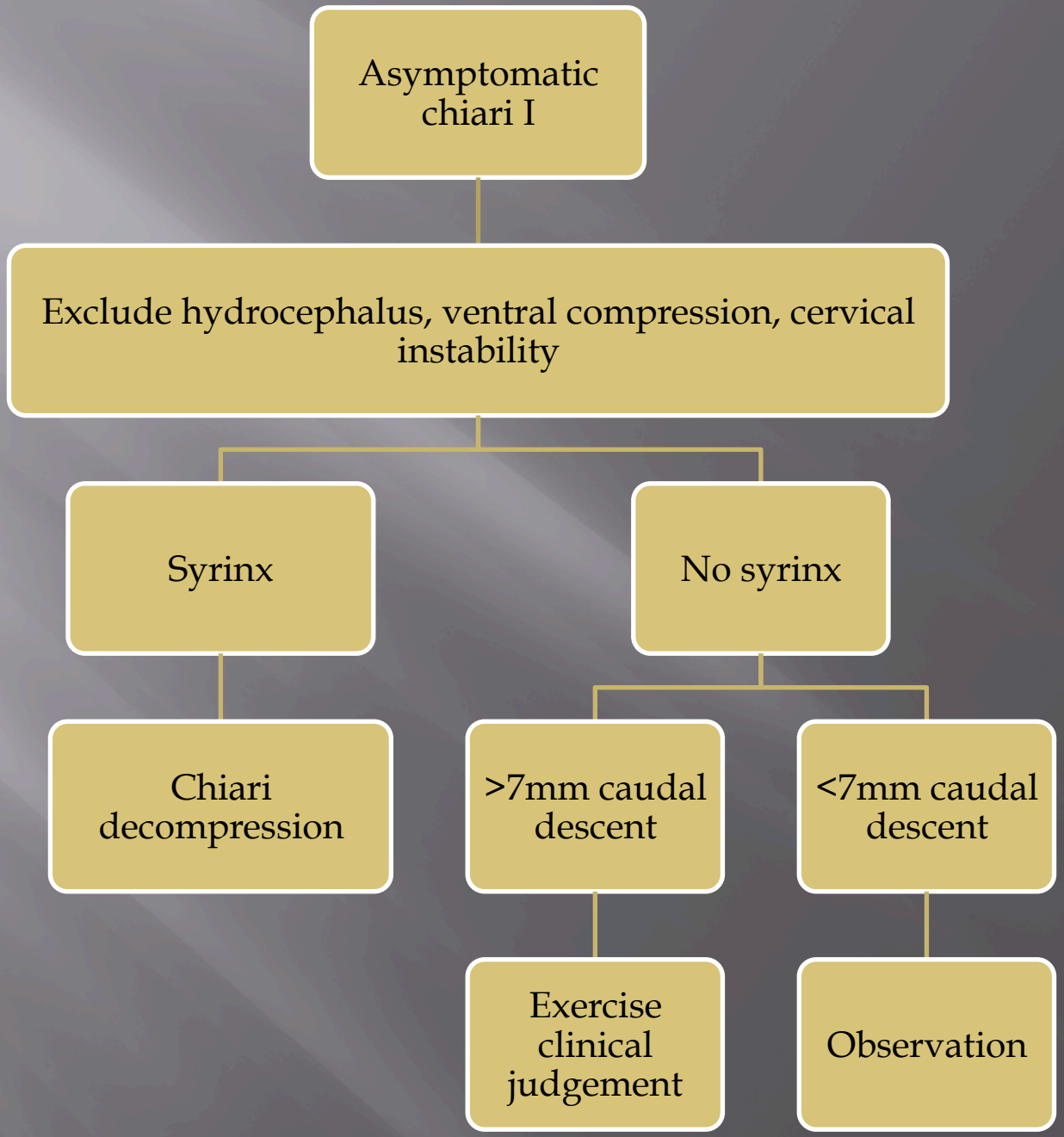
- Signs and symptoms related to syrinx
  - Dissociative sensory loss, upper limb weakness and thinning, lower limb spasticity.
  - Neuropathic pain in the extremities.
  - Neuropathic joints in upper extremities.
  - Uncommonly , JPS loss leading to sensory ataxia.
- Presentation usually in the 2nd and 3rd decades with a female preponderance

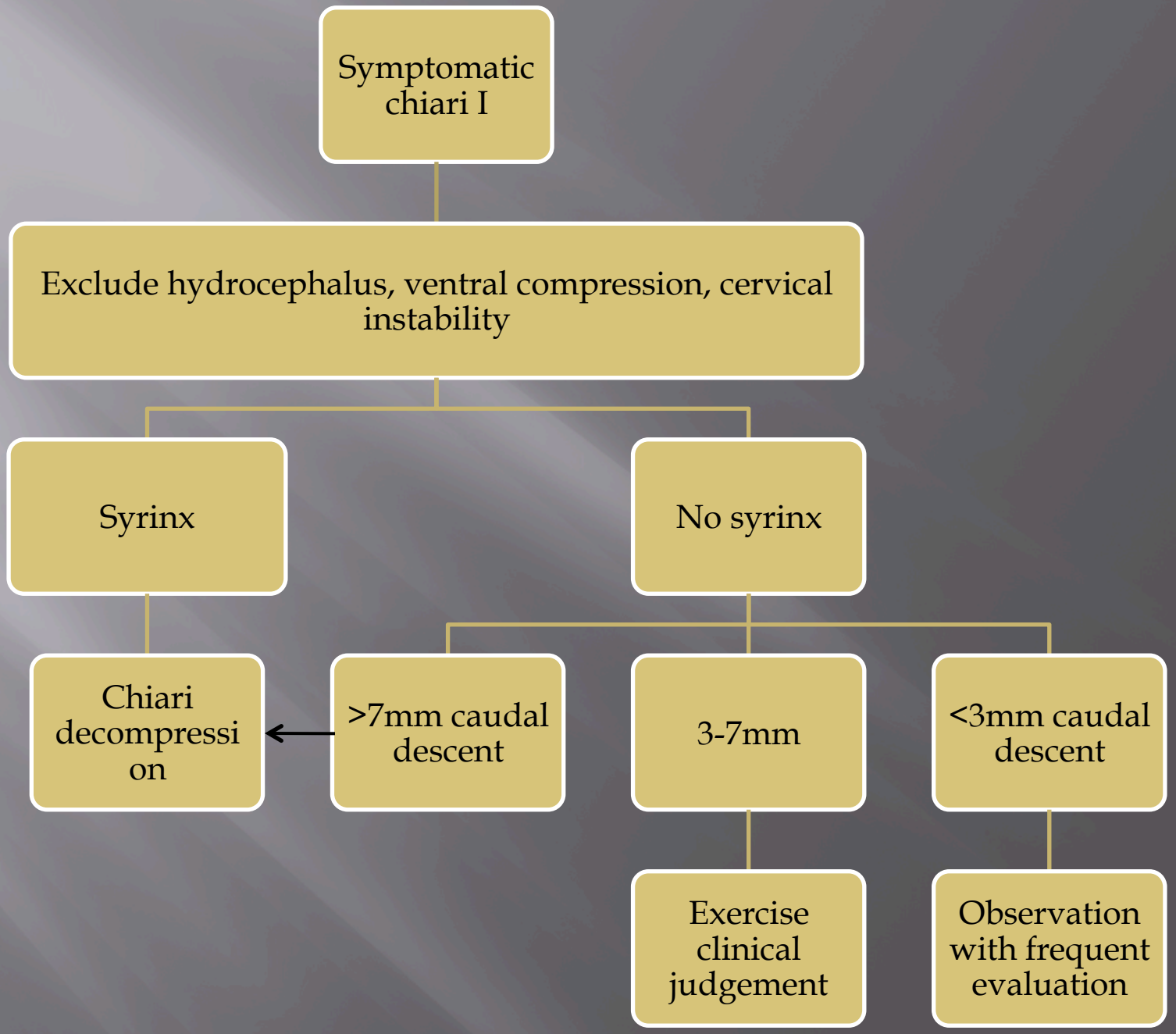
Theories explaining syringomyelia

- Gardner's hydrodynamic theory
  - Blocked fourth ventricular outlet
  - Pulsatile CSF pressure transmitted to central canal through obex "Water hammer"
- William's craniospinal dissociation theory
  - Valve like obstruction to free flow of CSF between cranial and spinal subarachnoid space at FM
  - Equalization of CSF pressure between cranial and spinal compartments hindered
  - CSF sucked into the syrinx.

- Oldfield's theory
  - Systolic downward motion of the tonsils creates a piston effect on the cervical spinal cord
  - Interstitial fluid driven into the central canal distending it.
- *No theory however, successfully explains all observations*

Approach to treatment of Chiari I


Asymptomatic chiari I
Exclude hydrocephalus, ventral compression, cervical instability
Syrinx
No syrinx
Chiari decompression
>7mm caudal descent
<7mm caudal descent
Exercise clinical judgement
Observation


Symptomatic chiari I
Exclude hydrocephalus, ventral compression, cervical instability
Syrinx
No syrinx
Chiari decompression
>7mm caudal descent
3-7mm
<3mm caudal descent
Exercise clinical judgement
Observation with frequent evaluation

Chiari I decompression

- Aim of the surgical procedure
  - Establishment of normal CSF outflow from the ventricles
  - Increasing posterior fossa volume

- Surgical options
  - Suboccipital bone removal
  - Dural opening with or without closure
  - Arachnoid opening and hitching
  - Tonsillar reduction and opening of fourth ventricular outlet
  - Fourth ventricular shunting

- Suboccipital bone removal + C1 laminectomy
  - 3 X 3 cm suboccipital craniectomy
  - Dura left intact
  - Dura inelastic hence, decompression inadequate
  - Larger craniectomies with dural opening result in cerebellar ptosis.
- Dural opening with or without closure
  - Options
    - Only superficial layer divided
    - Durotomy with intact arachnoid
    - Augmentation duraplasty
    - Williams procedure – dural edges sutured to the muscle
    - At craniocervical juction, division of thick dural band

- Opening the arachnoid
  - Required when significant tonsillar descent with syringomyelia
  - Arachnoid bands divided
  - Arachnoid pegged to the dural edges
  - Augmentation duraplasty

- Tonsil reduction
  - Subpial coagulation
  - Subpial resection when tonsils gliotic
- Fourth ventricular shunting
  - When tonsils encased in dense arachnoid scar
  - Shunt tubing inserted under USG guidance into the fourth ventricle and communicated to cervical subarachnoid space
- Obex plugging redundant

- Complications
  - Aseptic meningitis (most common)
  - Wound dehiscence, pseudomeningocele
  - CSF leak

Chiari II malformation

- Defining features
  - Caudal descent of vermis, fourth ventricle and brainstem
  - Almost always associated with hydrocephalus and associated anomalies
  - Seen in almost all patients with myelomeningocele
  - 0.02% of all births with female preponderance

Theories

- Chiari's theory
  - Hydrocephalus leading to secondary Chiari
  - 10-20 % may not have hydrocephalus
  - Associated anomalies not explained
  - Chairi II features precede hydrocephalus
  - Small posterior fossa, low lying tentorium, upward cerebellar herniation not explained

- Cleland's theory
  - Primary dysgenesis of the hindbrain
  - Fails to explain supratentorial anomalies
- Induced small posterior fossa
  - Due to CSF leaking out from the open spinal cord defect
  - Fails to explain associated anomalies

- Penfield's traction theory
  - Traction by tethering of cord at the site of myelomeningocele pulls the post fossa contents
  - Traction effect however dissipated four spinal levels rostral
  - Fails to explain associated cranial deformities

- Unified theory of Mclone and Knepper
  - Currently most accepted
  - Both the open neural tube defect and incomplete occlusion of central canal responsible
  - Temporary occlusion of the neural tube (Day 23-32) mandatory for upstream ventricular distension
  - Posterior fossa not fully developed due to inadequate ventricular distension
  - Rapid growth of hindbrain later leads to herniation

Radiological findings

- Skull
  - Luckenschadel/ craniolacunia
  - Frontal bone scalloping "lemon sign"
  - Scalloping of petrous bone and jugular tubercles
  - Concavity of the clivus
  - Low inion, small post fossa
  - Enlarged foramen magnum
  - Clival concavity
  - Basilar invagination and atlas assimilation

- Spine
  - Cervical spinal canal enlarged.
  - Scalloping of the odontoid process
  - Incomplete posterior arch of C1
  - Klippel-Feil deformity

- Ventricle and cistern
  - Hydrocephalus seen in 90%.
  - Fourth ventricular outlet obstruction responsible
  - Aqueductal stenosis uncommonly responsible for hydrocephalus
  - Medial pointing of the inferior margins of floor of lateral ventricles
  - “Colpocephaly”
  - Fourth ventricle typically small, flat and elongated
  - Lateral recesses not well defined

- Meninges
  - Tentorium cerebelli usually widened heart shaped
  - Low lying, hypoplastic
  - Falx cerebri fenestrated/hypoplastic

- Spinal cord
  - Myelomeningocele always associated with Chiari II
  - Syringomyelia in 20-95%
  - Shortened cervical cord
- Telencephalon
  - Complete partial agenesis of corpus callosum/ septum pellucidum
  - Polygyria
  - “Chinese lettering”- Interdigitation of occipital / parietal lobes

- Gray matter heterotopia
- Agenesis of olfactory tract/bulb/cingulate gyrus

- Diencephalon
  - Enlarged massa intermedia
- Mesencephalon
  - Tectal beaking due to fusion of the colliculi
  - Midbrain typically elongated
  - Cranial nerve nuclei may be malformed
  - Aqueduct may be stenotic, stretched, posteriorly kinked or forked

- Metencephalon
  - Cerebellum grossly smaller and may tower above tentorium
  - Cerebellum may be displaced laterally spreading around the brainstem – "banana sign"
  - Lateral cerebellar edges may touch brainstem and basilar artery – "cerebellar inversion"
  - Pons elongated and flattened

- Myelencephalon
  - Medullary kinking, elongation and flattening
  - Pyramidal decussation more cephalad than normal

Clinical presentation

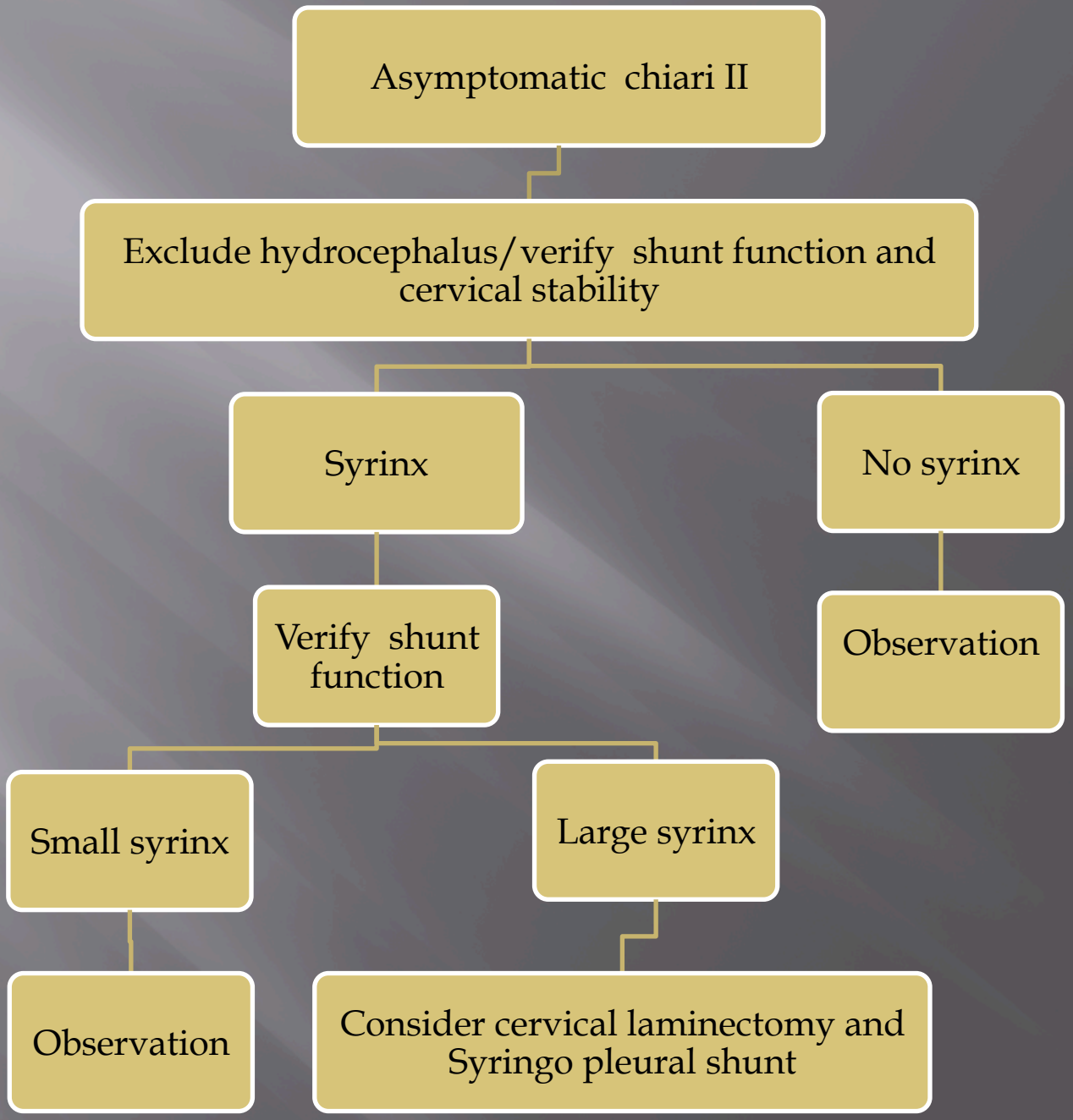
- Most common is with open neural tube defects
- Symptomatic chiari II is the most common cause of death in children <2 yrs of age with MMC.
- Symptomatic patients can be classified according to age at presentation
- Whatever be the age of child, hydrocephalus/ shunt malfunction should be excluded

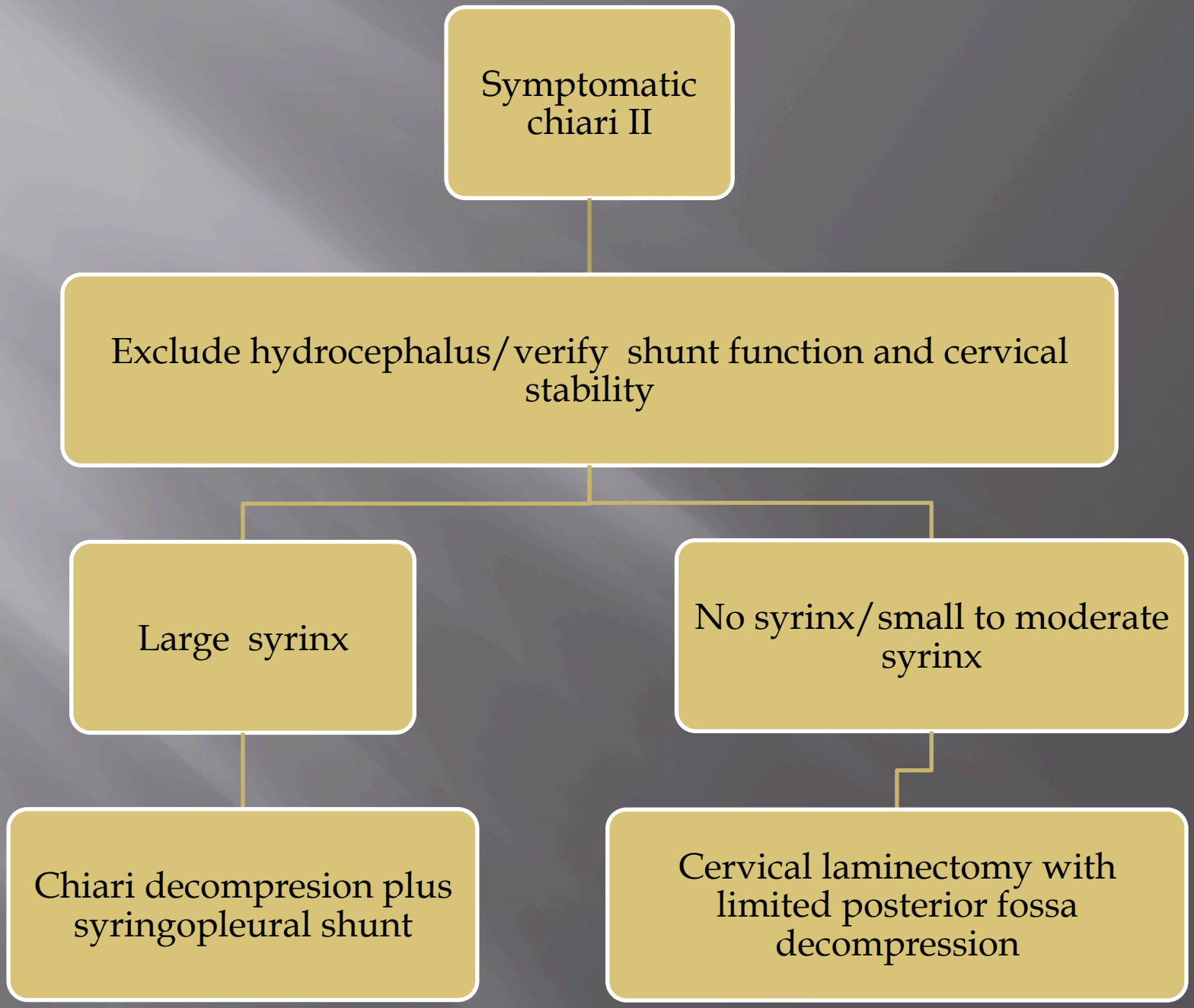
- Age at presentation less than 2yrs
  - Most frequent symptoms related to brain stem and cranial nerve dysfunction
  - Symptomatic Chiari is a neurosurgical emergency in this group.
  - Most commonly inspiratory stridor and PEAC (prolonged expiratory apnea and cyanosis)
  - PEAC = Apneic spell+ ophistotonic posturing and cyanosis
  - Laryngoscopy may reveal impaired vocal cord abduction
  - Downbeat nystagmus, fixed retrocollis

- Other signs and symptoms
  - Impairment of gag
  - Dysphagia, chronic aspiration, nasal regurgitation
  - Quadriparesis, nystagmus , developmental delay
  - Weak cry.

- Presentation in older age group
  - Less serious and rarely an emergency
  - Hallmark is cervical myelopathy
  - Weakness and spasticity in upper limbs
  - Suboccipital headache
  - Ataxia
  - Hand weakness , atrophy
  - Syringomyelia and associated symptoms
  - Ophthalmic problems common in adults

Chiari II
Approach to management


Asymptomatic chiari II
Exclude hydrocephalus/verify shunt function and cervical stability
Syrinx
No syrinx
Verify shunt function
Observation
Small syrinx
Large syrinx
Observation
Consider cervical laminectomy and Syringo pleural shunt


Symptomatic chiari II
Exclude hydrocephalus/verify shunt function and cervical stability
Large syrinx
No syrinx/small to moderate syrinx
Chiari decompresion plus syringopleural shunt
Cervical laminectomy with limited posterior fossa decompression

Chiari decompression

- Surgical technique basically similar to that in chiari-I
- Cervical laminectomy should expose the inferior margin of the herniated cerebellum
- Limited suboccipital craniectomy
- Constricting dural bands divided
- Key objective – finding the outlet of fourth ventricle
- Choroid plexus may serve as a guide

- Obstructive vermian tissue may be divided / perforated to encourage CSF flow out of the ventricles.
- Medullary kink not to be confused with vermis.
- Arachnoid clipped to dura with augmentation duraplasty
- Complications of this procedure similar to that for Chiari I.

Chiari III and IV

- Chiari III
  - Very rare
  - Occipital or cervical encephalocoele along with chiari II anomalies
- Chiari IV
  - No hindbrain herniation
  - Cerebellar hypoplasia or aplasia

Miscellaneous

- Chiari zero
  - CSF equilibrium changes at cranio-cervical junction
  - No hindbrain herniation
  - Syringohydromyelia
  - Post fossa decompression leads to dramatic improvement
  - Other causes of syrinx must be excluded.

Thank you