The Child With Cerebral Palsy

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Cerebral Palsy is not a disease

- It's a symptom complex or "group of disorders"
- Causing activity limitation
- Attributed to a non-progressive disturbance
- Occurred in the developing fetal or infant brain (in general the disturbance resulting in CP is presumed to occur before the affected function has developed.
- Often accompanied by disturbances in sensation, cognition, communication, perception, perception, orthopedic issues, and/or behavior, and/or by a seizure disorder.
- While the brain lesion is static, the course of the condition is not

The most common motor disability of childhood

- 3.6 per 1000 live births
- 10,000 new cases per year
- 80% antenatal factors

Most common motor disability of childhood

Spina Bifida	
Autism	
Brain Injury	
Cerebral palsy	

KEY FEATURES OF THE EPIDEMIOLOGY OF CP

- Found in about 1 in 500 school children
- Not as closely associated with “birth asphyxia” as once thought
- Increasing evidence for a role of antepartum infection
- Transient hypothyroxinemia may harmful?
- Lots of room for more research!

Epidemiology

- 5.2 per 1000 neonatal survivors at 12 months of age
- By age seven 2.5 per 1000 live births (but 10 times more likely to be retarded)
  - Nelson & Ellenberg *Pediatrics* 69:5, 1982
- 8000 new cases per year
- the overall prevalence of CP in 1-year survivors increased from 1975-1991 by 18%
- inflammatory cytokines released during the course of intrauterine infection play a central role in the genesis of preterm parturition, fetal PVL, and cerebral palsy
  - Yoon et al *BJOG*. 2003;110:124-127.

What causes CP?

We're not always sure……

- The rate of CP does not exceed 2% following placenta previa, abruptio placenta, breech, cord prolapse, nuchal cord, mid or high forceps delivery
- 75% of children with CP have normal Apgar scores
- Most children with an Apgar score of 3 at 10 and 15 minutes do <u>not</u> have CP

MAYBE IT'S BIRTH COMPLICATIONS?

DO COMMON LABOR COMPLICATIONS CAUSE CP?

	CP RISK
NO COMPLICATION	0.3%
NUCHAL CORD (18%)	0.3%
2ND STAGE ≥ 1 HOUR (10%)	0. 3%
MECONIUM (20%)	0.4%
MID OR HIGH FORCEPS (8%)	0.4%

Nelson KB, Ellenberg JH: *JAMA* 1984;251:1843-8

DO *UN*COMMON LABOR COMPLICATIONS CAUSE CP?

	CP RISK
◆ CORD PROLAPSE	0.3%
◆ PLACENTA PREVIA	0.6%
◆ BREECH POSITION	1. 0%
◆ ABRUPTIO PLACENTAE	1.9%

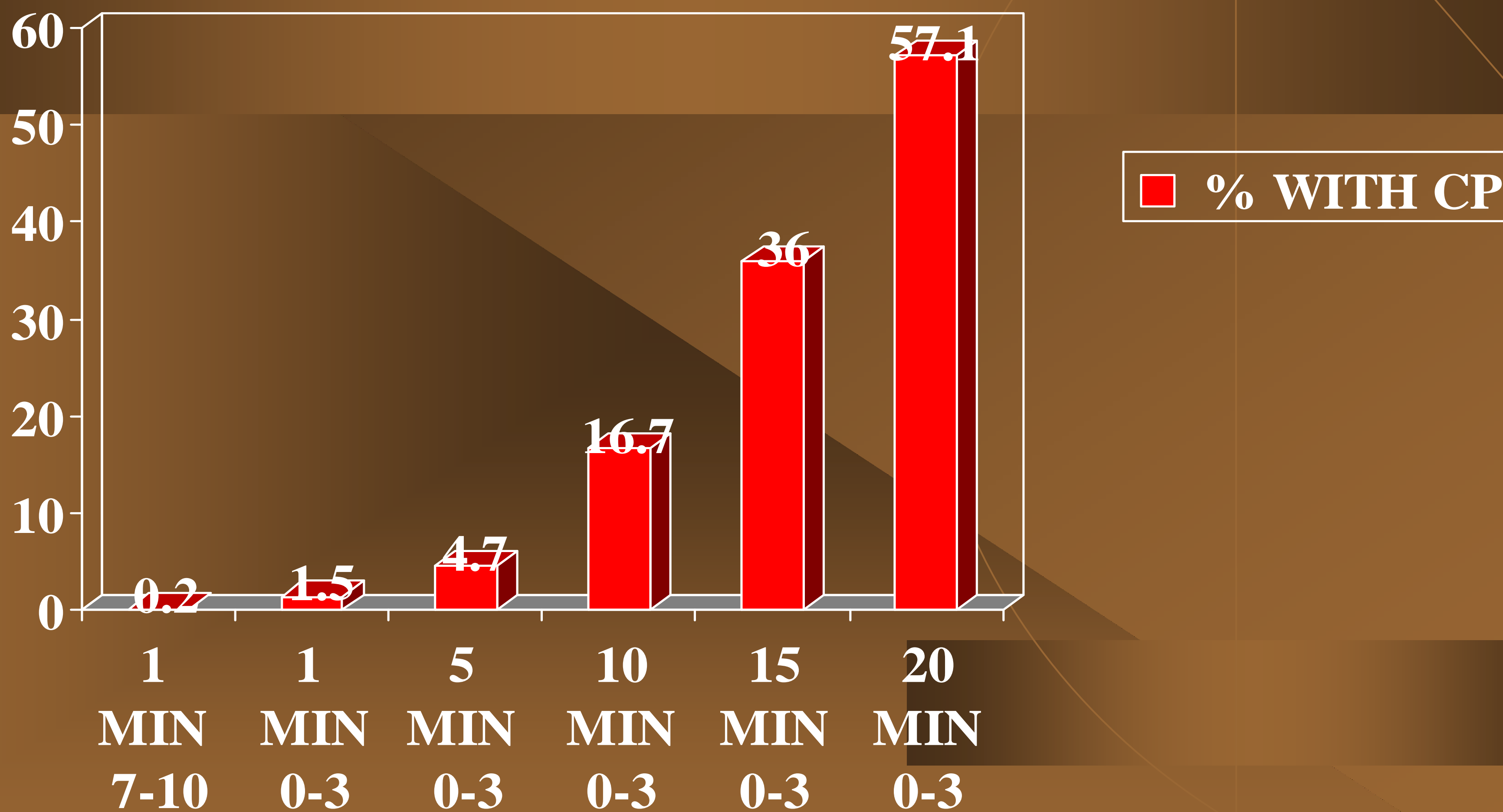
Nelson KB, Ellenberg JH: *JAMA* 1984;251:1843-8

DO LABOR COMPLICATIONS MATTER AT ALL TO CP RISK?

	CP RISK
◆ ***NO*** **LABOR COMPLICATIONS**	0.3%
◆ ***ANY*** **LABOR COMPLICATIONS**	0.3%

APGAR SCORES AND CEREBRAL PALSY

Source: Nelson KB, Ellenberg JH: Pediatrics 1979;64:225-32



WHAT ABOUT BIRTH ASPHYXIA CAUSING HYPOXIC ISCHEMIC ENCEPHALOPATHY (HIE)?

Not so much.....

- Intrapartum asphyxia: a rare cause of CP

  Blair & Stanley *J Peds* April 1988
- 1975-1980 All children with spastic CP born in Western Australia (n=183)
- Matched group of controls (n=549)
- Info on perinatal events was collected
- Birth asphyxia *was* associated with CP (RR 2.84; 95% confidence interval)
- BUT of all children with spastic CP intrapartum asphyxia was the possible cause of their brain damage in only 8% (15 of 183).

HOW ABOUT PREMATURITY?

PREMATURITY IS THE MOST IMPORTANT KNOWN ANTECEDENT

- 40-fold higher risk in infants < 1,500 g
- Possibly increasing in prevalence due to increased VLBW survival
- IVH (grades III and IV) is an important cause
- The brain lesion resulting after IVH is periventricular leukomalacia (PVL)
- PVL is the most important identifiable risk factor in the development of CP

BUT MOST KIDS ARE BORN AT TERM
WITHOUT COMPLICATIONS.

Risk of CP and gestational age

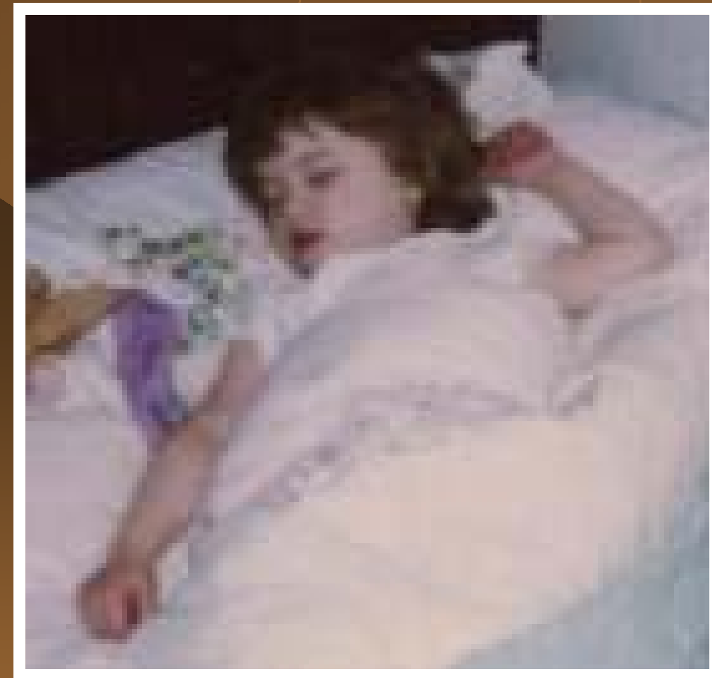
But there *are* some risk factors

- Low birth weight
- prematurity (<1500 grams)
- Maternal MR
- Maternal sz
- Previous pregnancy loss
- Multiple pregnancies
- Motor deficit in an older sibling
- Male gender
- Infections (rubella, toxoplasmosis, CMV, etc)
- Intrauterine infection w/chorioamnionitis
- Jaundice (kernicterus)
- Asphyxia
- Maternal thyroid deficiency *(Bernal and Nunez 1995).*
- Non-vertex presentation
- fetal inflammatory response has been found to be related to white matter injury and CP

Diagnostic clues but certainties

- Failure to meet expected developmental milestones
- Failure to suppress obligatory primitive reflexes.
- Abnormal muscle tone: hypotonic / hypertonic. Often will have early hypotonia followed by hypertonia as the brain myelinates.
- Definite hand preference before age 1 year
- Asymmetric crawling or failure to crawl
- Growth disturbance especially failure to thrive
- Persistent primitive reflexes

Persistent primitive reflexes - ATNR



Persistent Primitive Reflexes - Moro

Differential

- Hereditary spastic paraplegia
- Rett syndrome
- Tethered spinal cord
- Genetic d/o (named and unnamed)
  - Brown Vialetto VanLaer ?
  - Aicardi
- Charcot Marie Tooth Disease
- Spinal Muscular Atrophy
- Spina Bifida
- Polio
- Myopathy

Mean Age for Diagnosis

- Quadriplegia - 5 months
- Diplegia - 12.5 months
- Hemiplegia - 21 months

Classification based on topography

- Monoplegia: one limb involved
- Diplegia: primarily legs are involved
- Hemiplegia: one side of body involved
- Quadriplegia: all four limbs and trunk involved

Classification based on tone disorder

- Spasticity: velocity-dependent increase in tone
- Athetosis: slow, writhing, involuntary movements especially in distal extremities
- Chorea: abrupt, irregular, jerky movements usually in the head, neck, and extremities
- Dystonia: slow, rhythmic movements with tone changes generally found in the trunk and extremities
- Ataxia: unsteadiness with uncoordinated movements;   wide-based gait

Associated Deficits

- Mental retardation: 30-50%
- Seizures: half of all CP, but 70% of hemiplegics
- Oromotor: sucking, swallowing, excessive drooling, articulating, poor dentition, FTT
- Pulmonary - aspiration
- GI: reflux, constipation
- Ocular/Visual: strabismus, refractory errors
- Perceptual deficits
- behavioral disorders
- bladder/bowel control

Is this CP?

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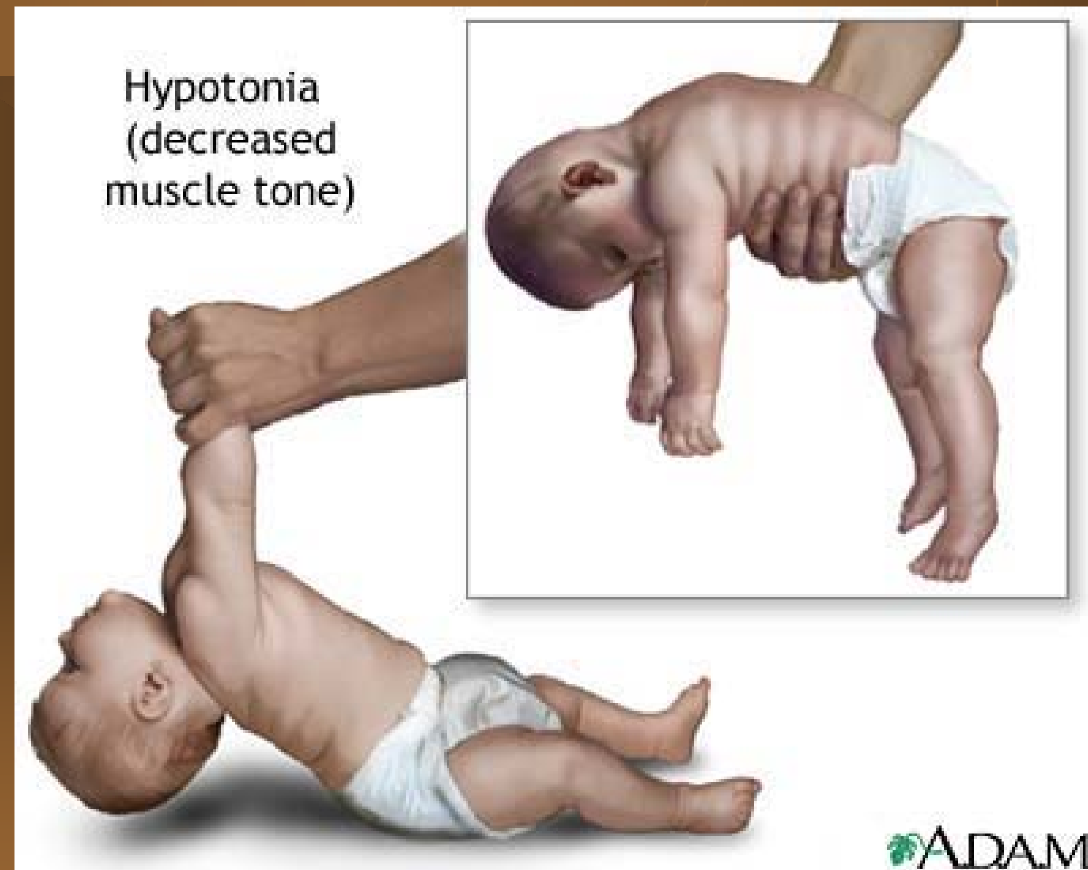

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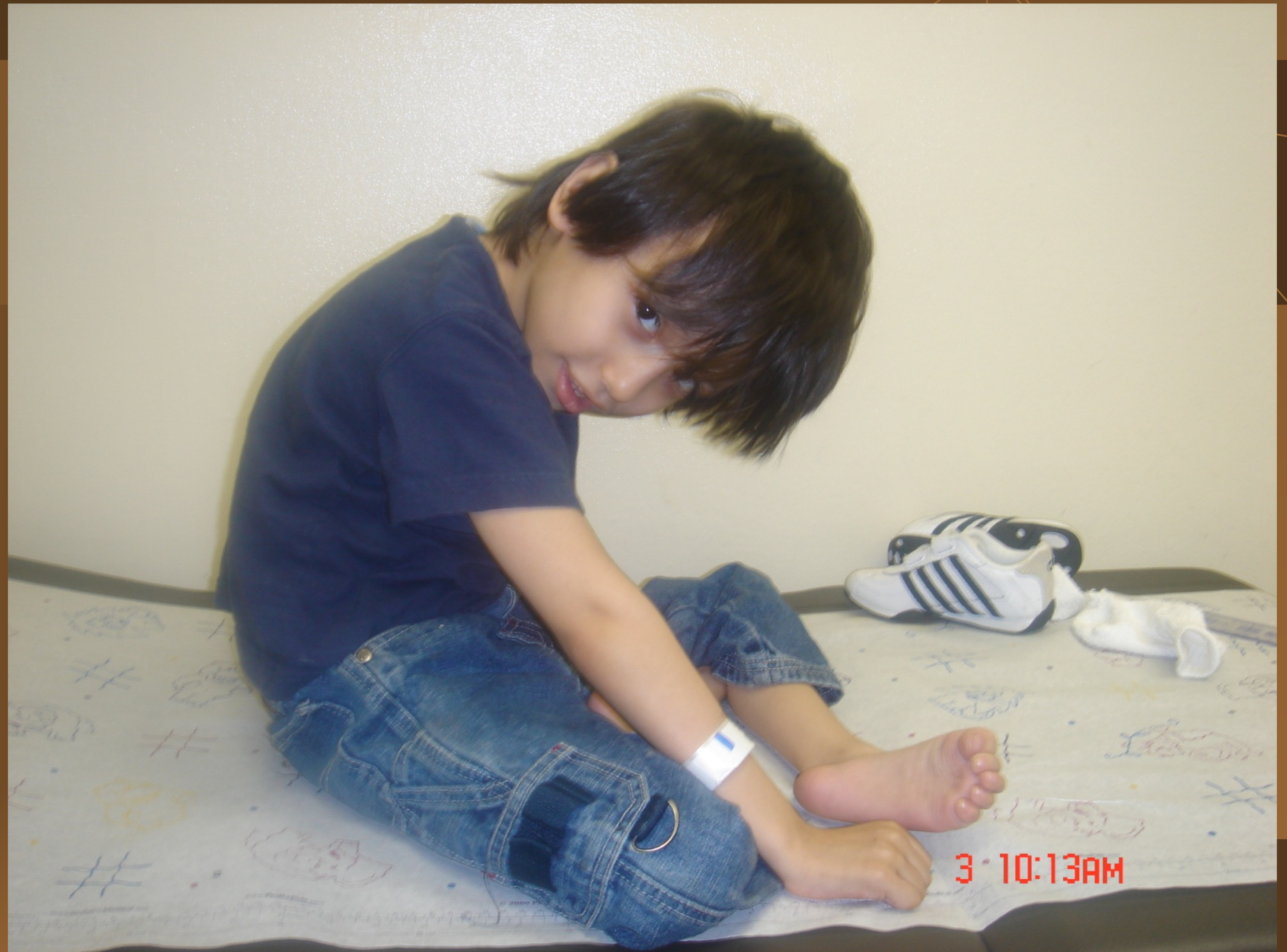


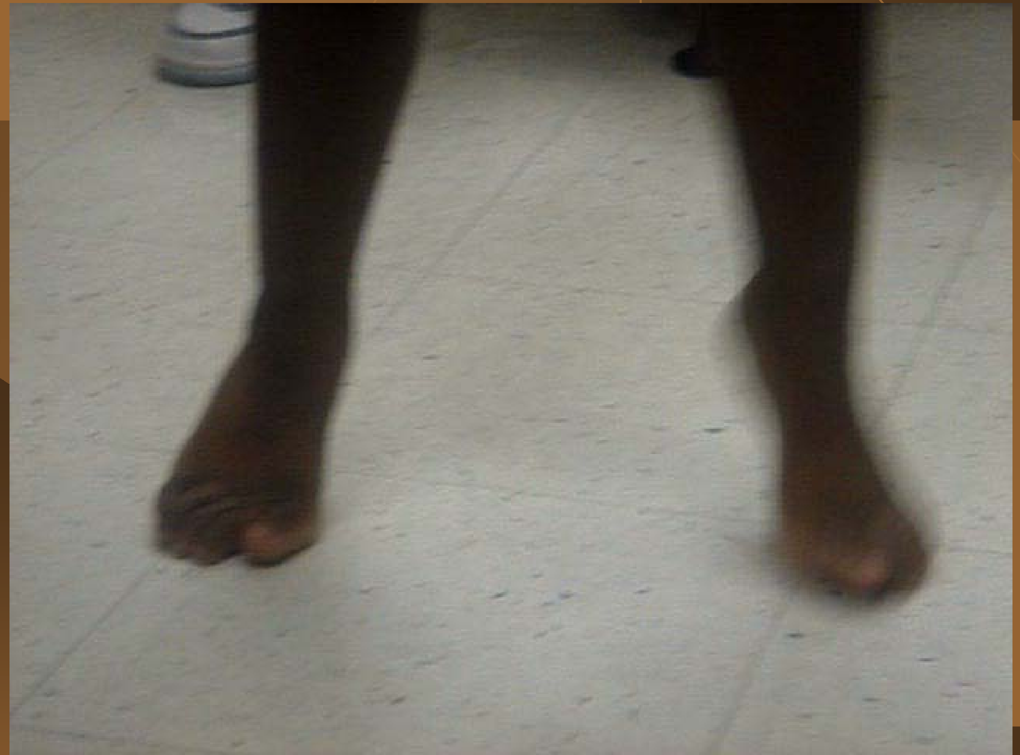
Do Musculoskeletal Complications Help in Dx?

- Caused by imbalance between agonists and antagonists
- muscles in a growing child tend to shorten if not fully stretched daily
- Progression may be slowed by consistent proper positioning

Do Contractures Help in Dx?

How about spasticity?

Is this CP?



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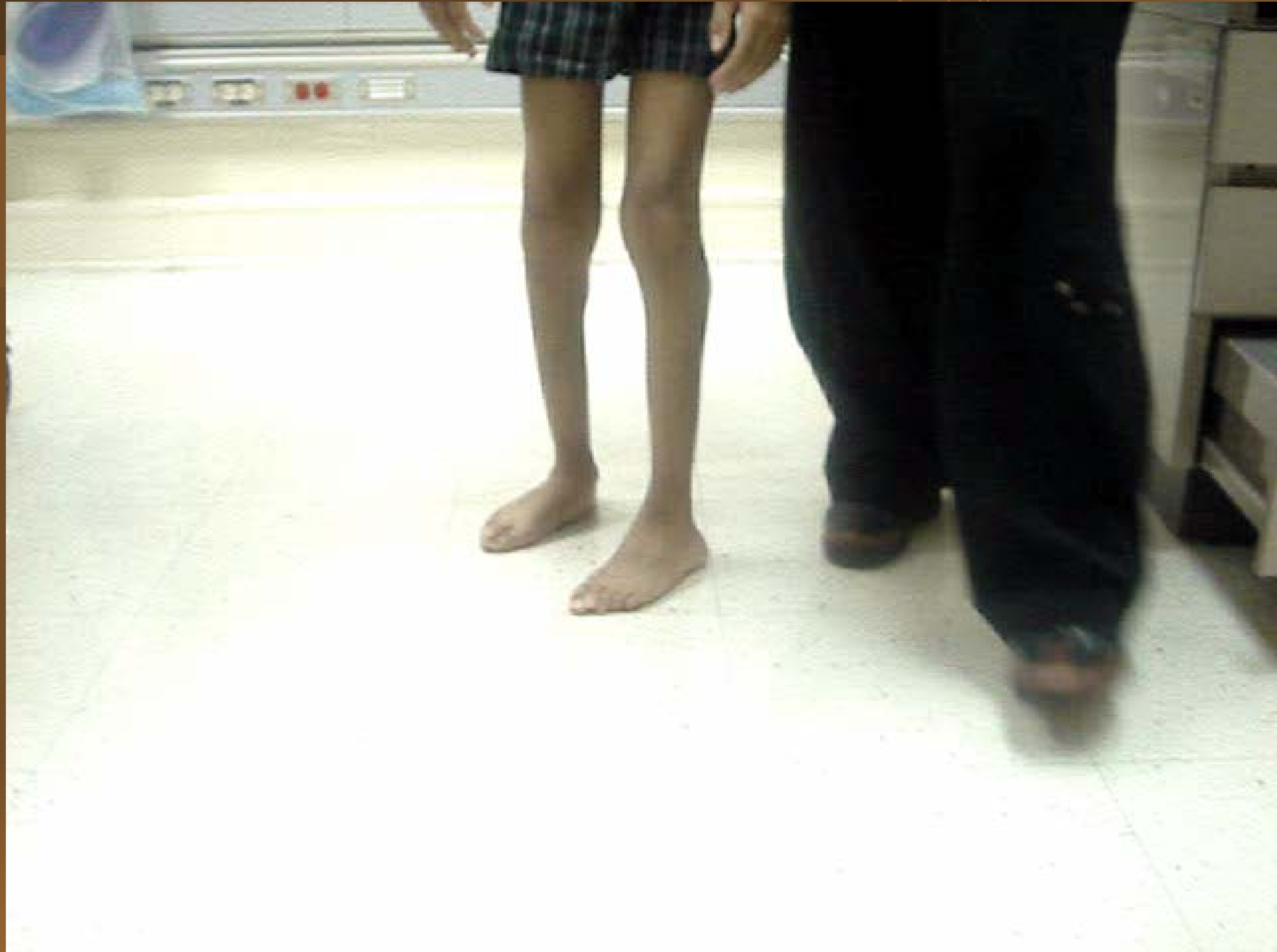


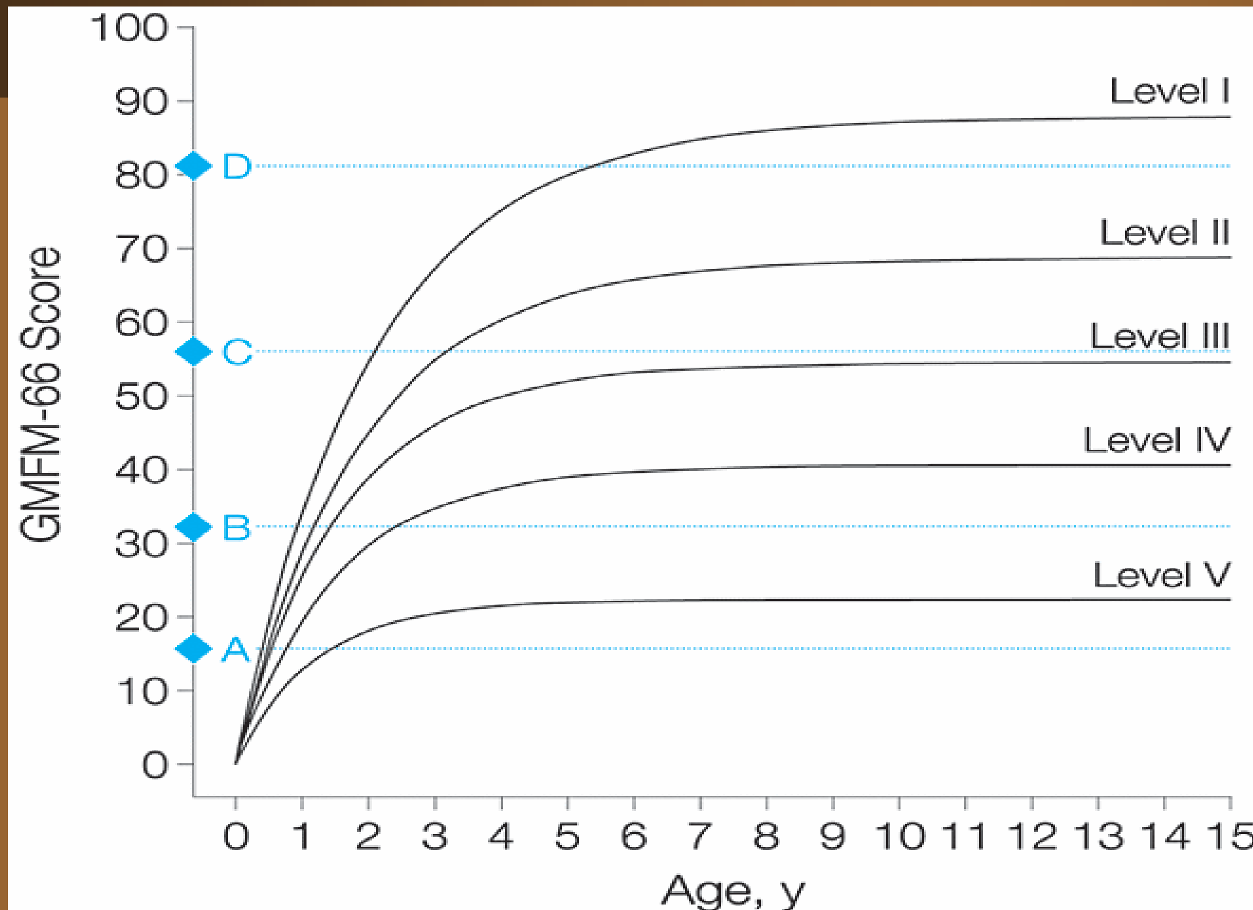
Diagnostic Testing

- Neuroimaging (MRI preferred) to determine the presence and nature of any brain abnormality (normal results do not exclude CP)
- Possible genetic evaluation to assure that the condition is not progressive
- Because of the high incidence of associated conditions screen for:
  - mental retardation
  - ophthalmologic disorders
  - hearing impairments
  - Speech and language disorders

What's not CP?

- Anything degenerative
- If there are no tone abnormalities (idiopathic toe walking)
- If it's a genetic syndrome (Aicardi syndrome, Palizaeus-Merzbacher)
- If it's not caused by a brain lesion (spina bifida or spinal cord injury)

Children, on average, reach about 90% of their motor function (as measured by the GMFM-66) by around age 5 years or younger, depending on their GMFCS level.



- A: lift and maintain head in a vertical position with trunk support by a therapist while sitting;
- B: when in a sitting position on a mat, a child can maintain sitting unsupported by his/her arms for 3 seconds;
- C: ability to walk forward 10 steps unsupported;
- D: walking down 4 steps alternating feet with arms free.

Rosenbaum, P. L. et al. JAMA 2002;288:1357-1363.

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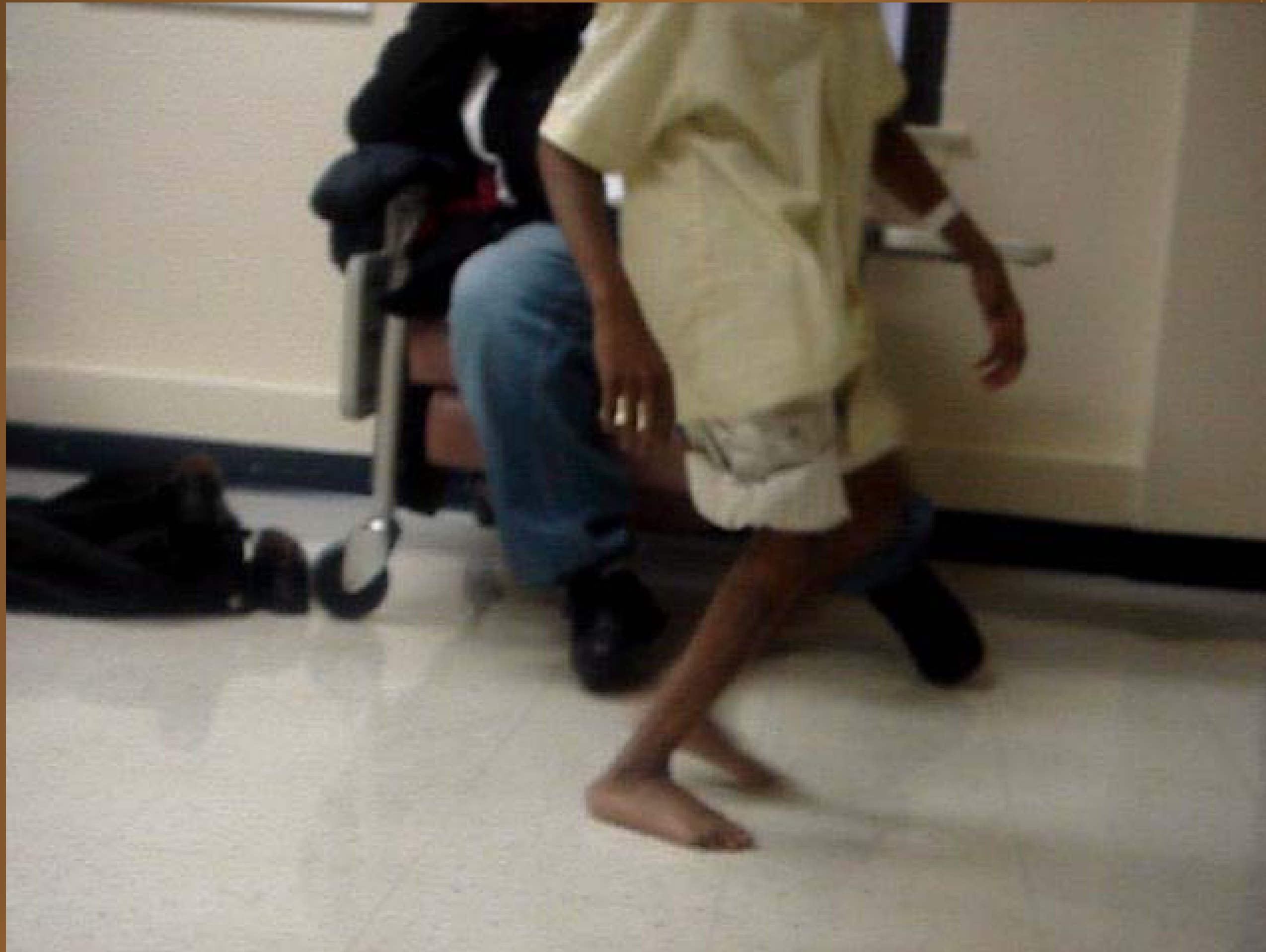

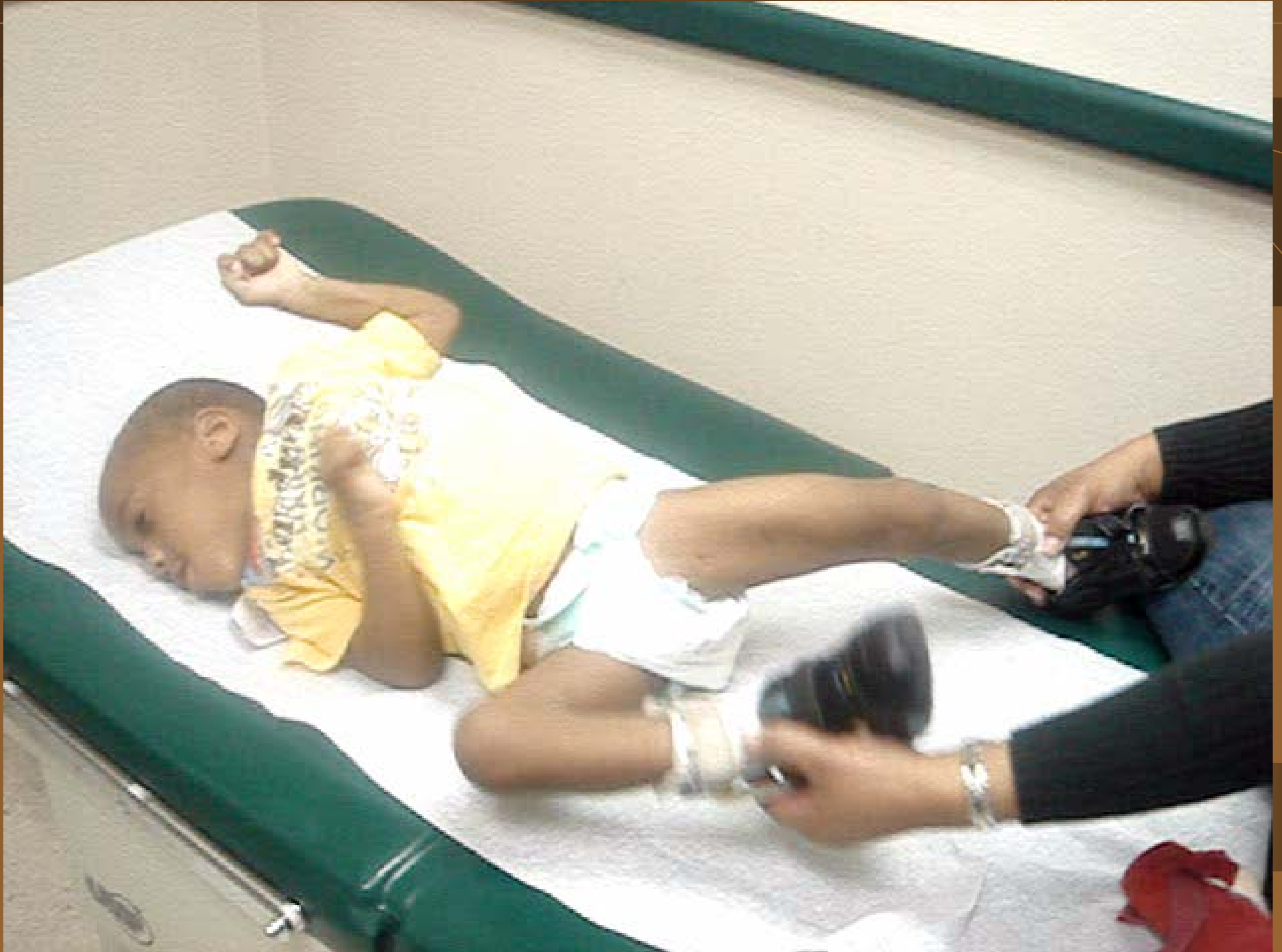

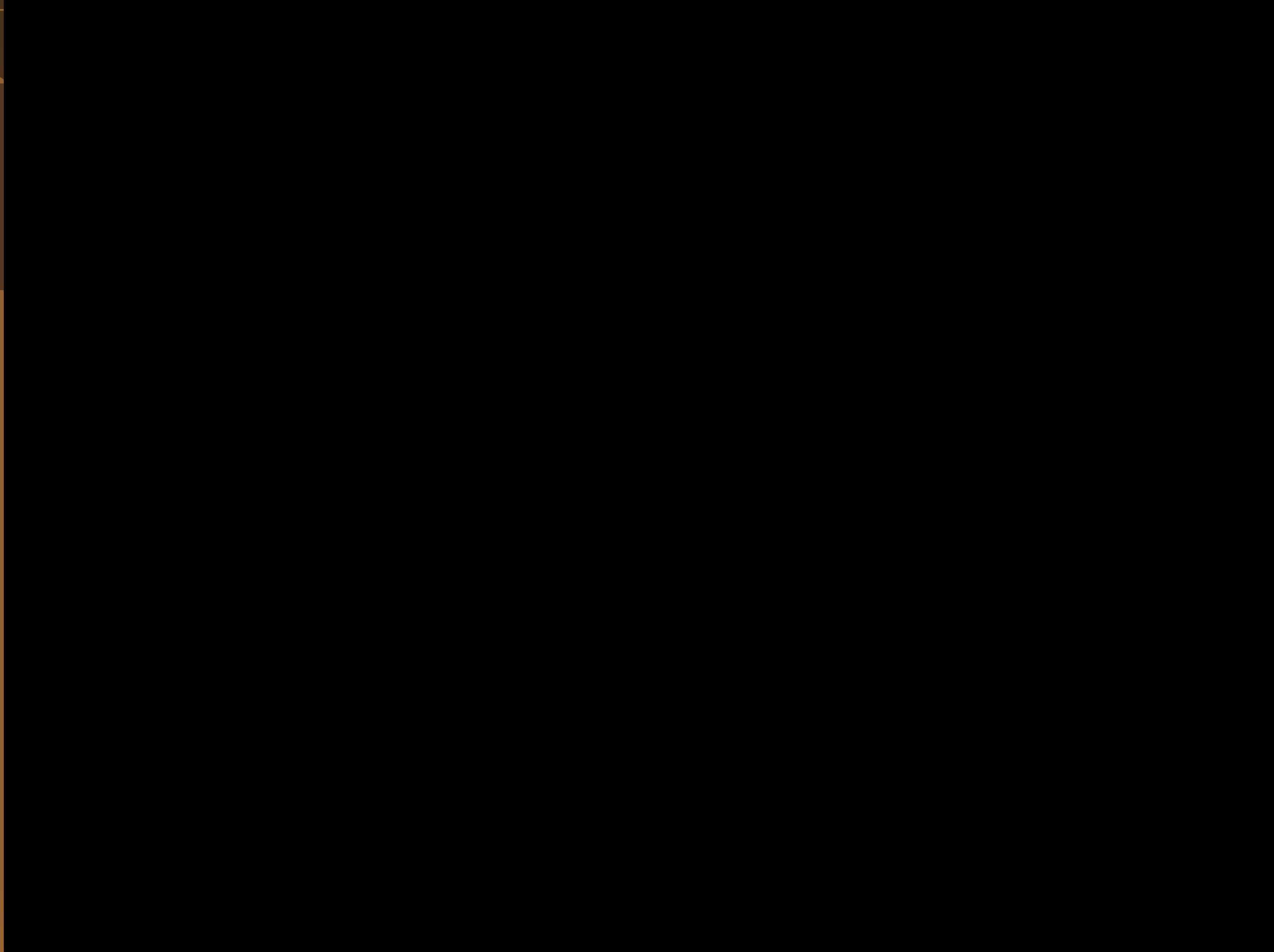

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