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Cerebral palsy

Fetal Alcohol Spectrum Disorders

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Cerebral palsy

Aims

1. Know the definition of CP.
2. Recognize the classifications of CP.
3. Know the basic therapeutic approach to CP.

CP definition

- CP is defined as a group of disorders resulting from permanent non progressive cerebral dysfunction developing before maturation of CNS affecting the locomotor system
- Cerebral palsy is a static encephalopathy (=brain injury), non progressive disorder of posture and movement
- Variable etiologies

CP definition

1. Disorder of movement and posture
2. Damage to immature brain
3. Permanent
4. Non progressive (static encephalopathy)
5. Non hereditary
6. Onset before or at birth or during early months of life.

- Although CP is „static”, non-progressive disorder, clinical picture may change with time.
- Contractures and postural deformities may become more severe with time or may improve with therapy.

Epidemiology

- The most common childhood movement disorder
- Prevalence of at least 1 to 2 cases per 1000 live births
- The incidence is higher in males.

Etiology

Prenatal (70-75%)

Perinatal (5-10%)

Postnatal

Etiology

Prenatal (70-75%)

- Congenital infections (toxoplasmosis, rubella, CMV, HSV)
- Fetal anoxia
- Toxic (nicotine, alcohol, drugs)
- Vascular
- Rh incompatibility
- Genetic
- Congenital malformation of CNS
- Exposure to radiation
- Diabetes
- EPH gestosis

Etiology

Perinatal (5-10%)

- Prematurity
- Low birth weight
- Birth asphyxia
- Traumatic / rapid / breech delivery
- Intracranial hemorrhage
- Metabolic (hypoglycemia)
- Meconium aspiration syndrome
- Excessive neonatal jaundice (kernicterus)

Etiology

Postnatal

- Cerfebral trauma
- Cerebral hemorrhage
- Neuroinfection (meningitis, encephalitis)
- Toxic
- Child abuse

In the vast majority of children with CP, the risk factors nor etiological factors can not be found.

Classification of CP

depending on tone or movement patterns

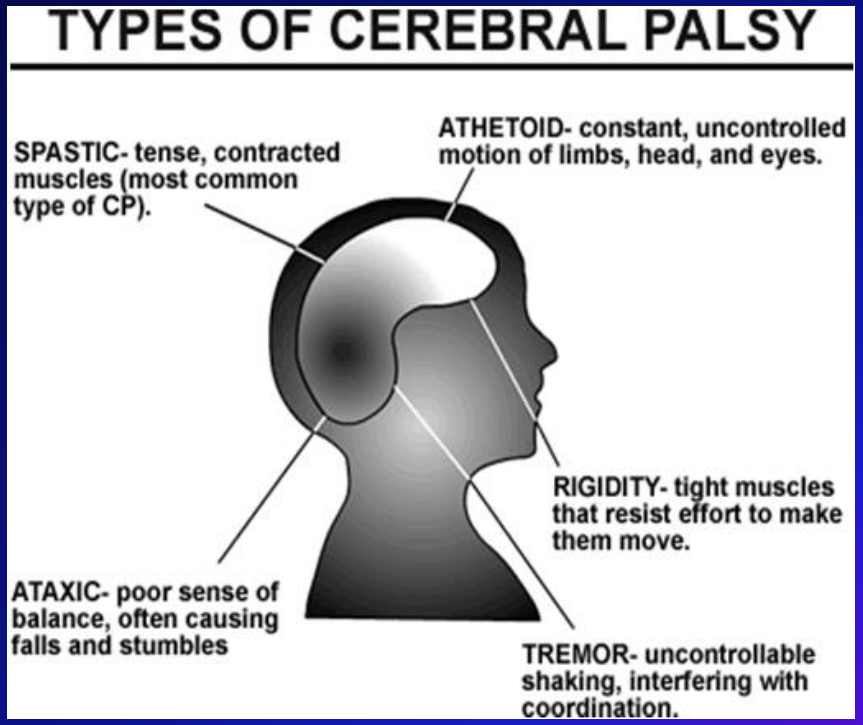
Pyramidal (70-80% of CP)

- Spastic (rigid)

Extrapyramidal

- Athetoid (dyskinetic)
- Ataxic
- Atonic (flaccid, hypotonic)

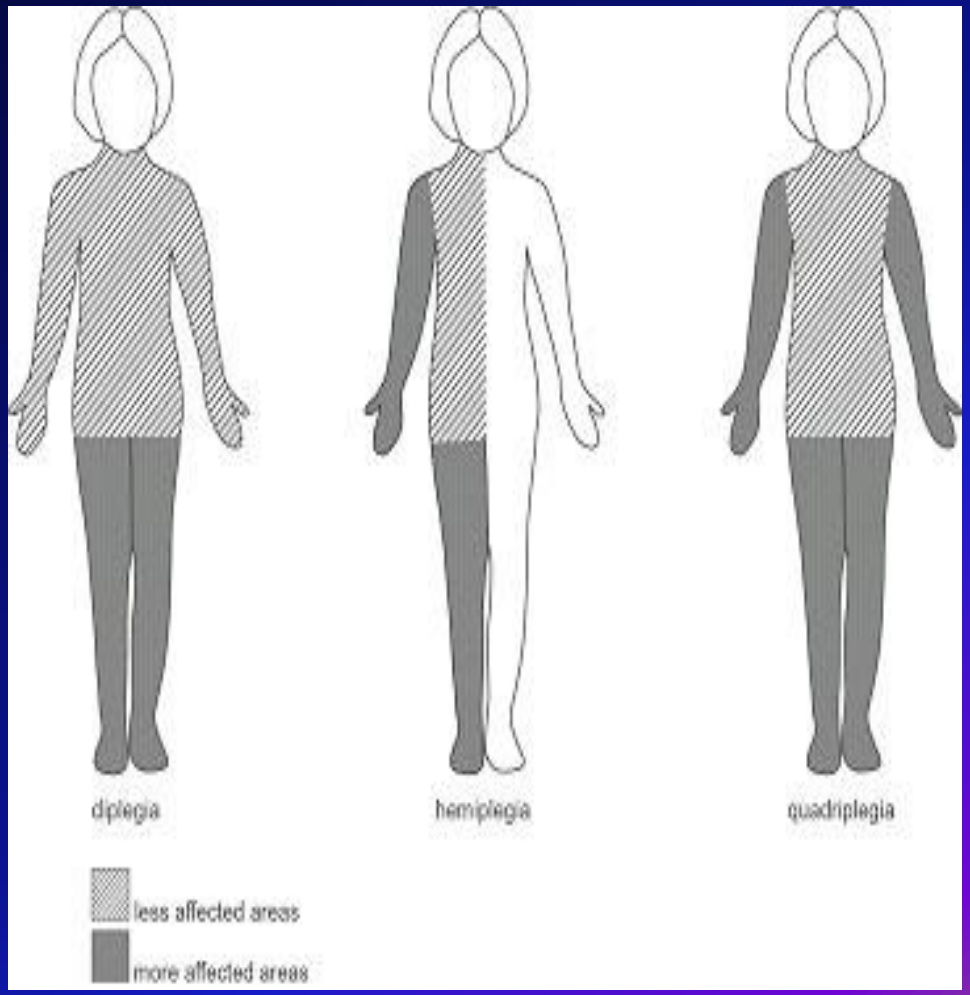
Mixed



Classification of CP

depending on the topographical distribution

- Monoplegic
- Diplegic / Paraplegic – the most common form
- Triplegic
- Hemiplegic
- Quadri(Tetra)plegic (double hemiplegia)



Quadri(Tetra)plegic, or double hemiplegia

The most severe form:

- Spactic paresis of all four limbs
- Pseudobulbar palsy
- Visual problems
- Mental impairment
- Epilepsy

Diplegic (paraplegic) form

- Spastic paresis predominantly of the lower limbs

  (upper limbs may be affected)
- Strabismus may be present
- Hearing, visual, or speech problems.
- Intellectual development within normal range

Diplegic (paraplegic) form

Ataxic CP (Cerebellar form)

- Muscular hypotonia
- Delays in posture and locomotion development
- Discoordination of the movements
- Speech problems, dysarthria
- Intellectual impairment is usually mild or moderate
- Often hydrocephaly

Dyskinetic cerebral palsy

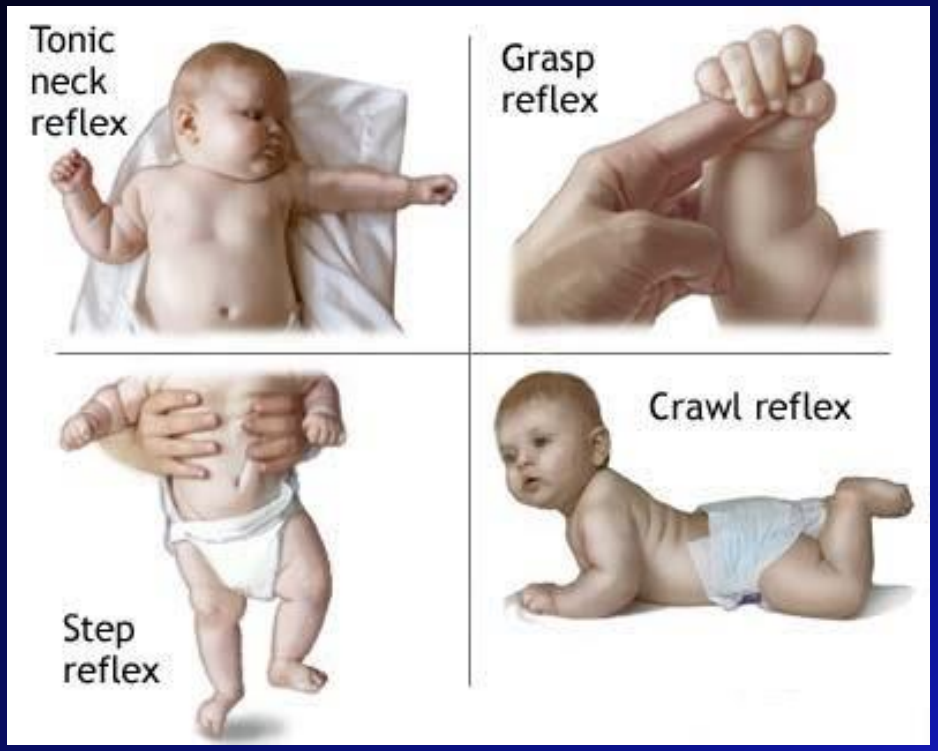
- Damage to the basal ganglia and the substantia nigra
- Occur during brain development due to bilirubin encephalopathy and hypoxic-ischemic brain injury
- Characterized by both hypertonia and hypotonia
- Inability to control muscle tone.
- An extrapyramidal form of cerebral palsy
- Two different groups: Choreo-athetotic CP is characterized by involuntary movements and tics, whereas dystonic CP is characterized by slow, strong contractions, which may occur locally or encompass the whole body

Clinical presentation may include:

- Manifestation may not be apparent before one year of age (usually around the time when the child starts to crawl).
- Delayed motor milestones
- Decreased muscular strenght
- Abnormality of muscular tone (hyper/hypotonia)
- Abnormal persistence of primitive reflexes (*Moro reflex)*

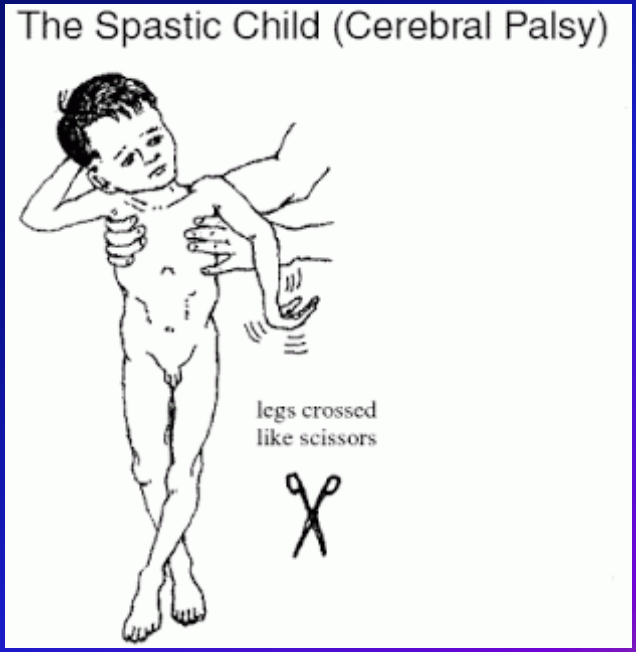
Clinical presentation may include:

- Abnormal persistence of primitive neonatal reflexes: Moro reflex >6mth, Step and grasp reflexes >3mth, Asymmetric tonic neck reflex >6mth)



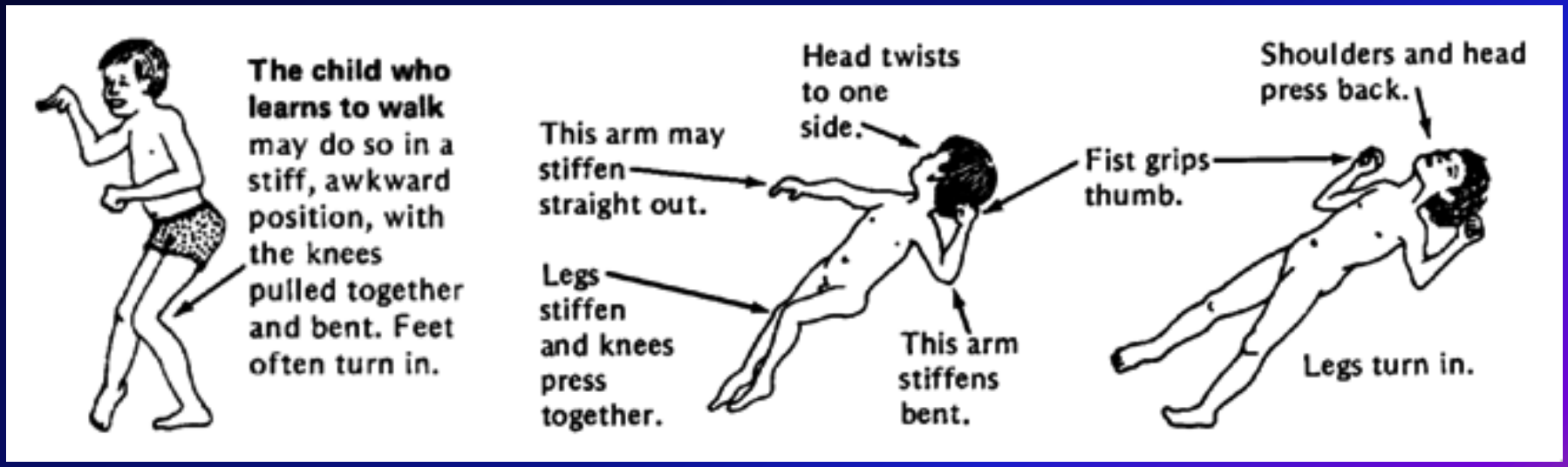
Clinical presentation may include:

- Excessive drooling
- Microcephaly (25%)
- Brisk reflexes
- Babinski sign positive after the age of 2 yrs
- Scissoring posture of legs
- Commando crawl



Clinical presentation may include:

- Tiptoe walking, spastic gait
- Paucity of movements and facial expression
- Arching back
- Clenching of hands with adducted thumb



Complications and associated deficits

- Seizure disorders 30-50%
- Mental retardation 50%
- Visual defects 50%
- Microcephaly 25%
- Hearing defects 15%
- Speech disorders 15%
- Psychological problems

Complications and associated deficits

- Digestive tract dysfunction: sucking and swallowing problems, dysphagia, gastroesophageal reflux disease (GERD), constipation.
- Kiphoscoliosis, hip joints dislocations
- Recurrent URTI, LRTI, and UTI

Diagnosis of CP

- Very difficult in a neonate/young infant.
- If a CNS injury is suspected, head imaging (by ultrasound or MRI) can be helpful in recognizing CP early.
- Possible imaging findings include periventricular leucomalacia, atrophy, or focal infarctions.
- Beyond infancy, CP is suspected when a child fails to meet anticipated developmental milestones.

Management

- CP cannot be cured.
- Depends on the pattern of dysfunction.
- Improve quality of life, functional abilities and participation in society.
- Treatment of concomitant disorders.

Management

- Pharmacological
  - Neurological: anticonvulsants, levodopa, baclofen, botulin toxin
- Surgical
  - joint & tendon release, skeletal deformation corrections
- Rehabilitative:
  - Occupational (OT) and physical therapy (PT)
  - Speech therapy
  - Adaptive equipment

Summary

- Cerebral palsy is a disorder of movement or posture resulting from an insult to, or an anomaly of, the central nervous system.
- Most children with cerebral palsy have no identifiable risk factors for the disorder.
- Optimal treatment plans for cerebral palsy use a multidisciplinary approach.

case

- A 12-month-old boy arrives for a wellchild visit.
- The mother is concerned that the baby's manner of crawling, where he drags his legs rather than using a fourlimbed movement, is abnormal.

- She says that the child only recently began crawling and he does not pull to a stand.
- You noted at his 6-month visit that he was not yet rolling over nor sitting; previous visits were unremarkable as was the mother's pregnancy and vaginal delivery.
- On examination today, you note that he positions his legs in a "scissoring" posture when held by the axillae.

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- What is the most likely diagnosis?
- What is the next step in the evaluation?

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- Gather detailed history, focusing on developmental questions; obtain a thorough pregnancy, birth, social, and family histories; and perform a detailed neurologic examination.

- What is the initial step in the evaluation of this child?
- What is the most likely diagnosis?
- What is the next step in the evaluation?

Spastic cerebral palsy (CP)

- What is the initial step in the evaluation of this child?
- What is the most likely diagnosis?
- What is the next step in the evaluation?

- Vision and hearing testing
- Consider a brain MRI scan
- Arrange for therapy with a developmental specialist / physiotherapist.

FASD

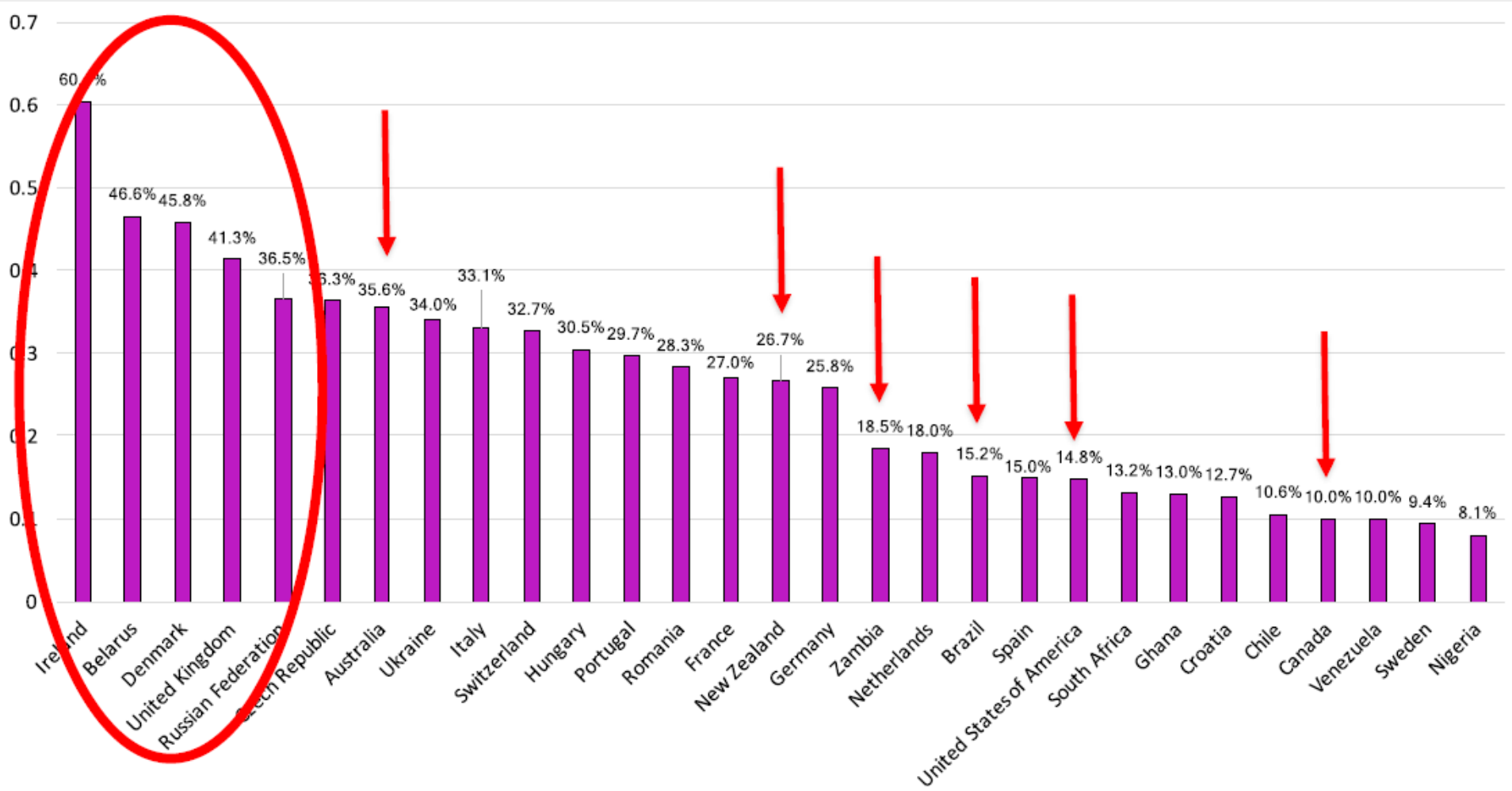
Fetal Alcohol Spectrum Disorders

The issues are not obvious

Maternal alcohol consumption is a significant public health concern worldwide.

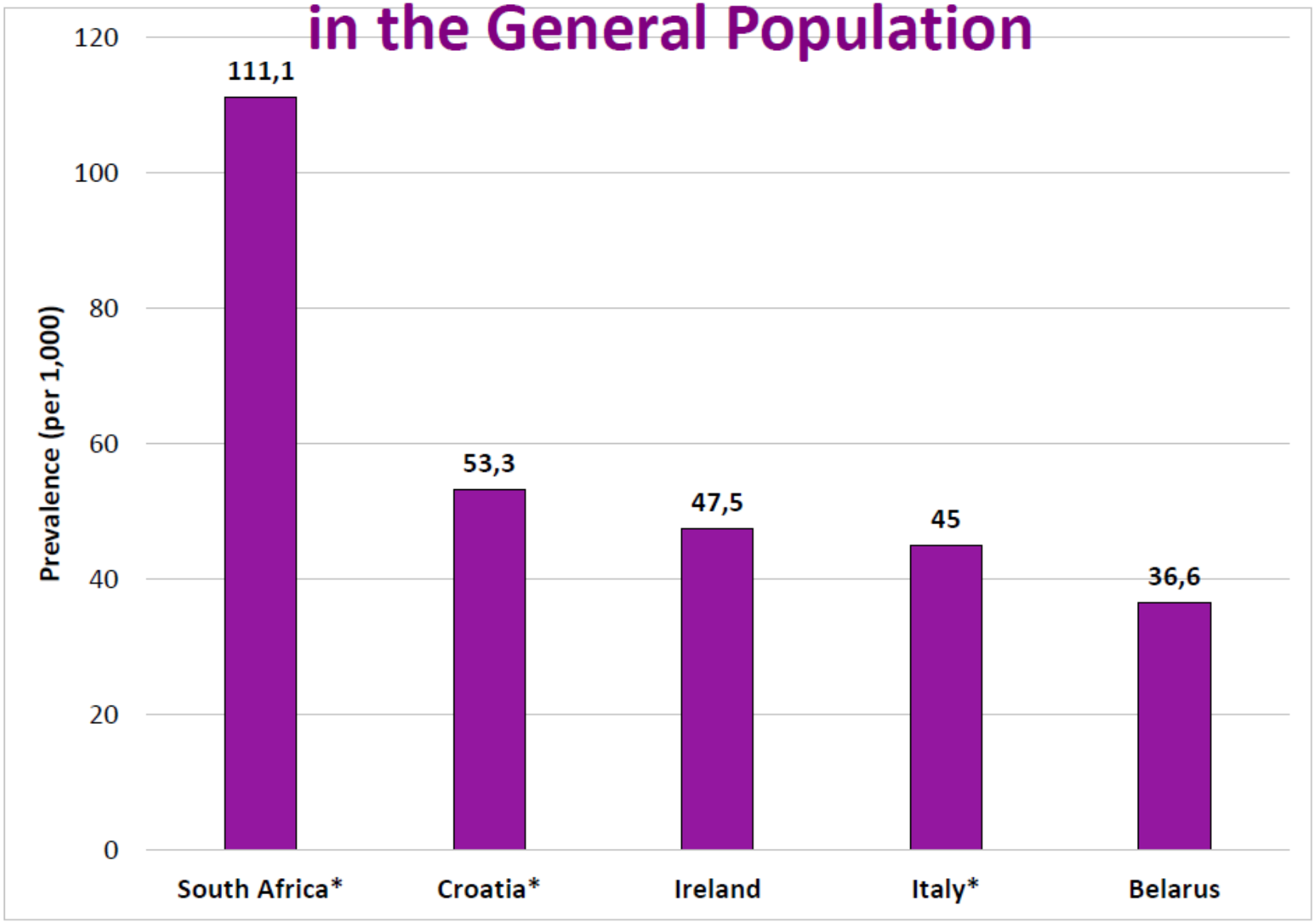
Prevalence of Alcohol Use during Pregnancy in General Population for select Countries

(any amount of alcohol consumed and at any point during pregnancy)



Popova et al., Lancet Global Health, 2017

Five Countries with the Highest Prevalence of FASD in the General Population



*Based on actual data (meta-analysis)

In Poland, about 9 000 children with FASD are born every year.

The prevalence of FASD in Poland has been reported as 20 per 1000 children, and the rate of complete FAS as 4 per 1000.

In the United States, those rates of FASD / FAS are 9 and 1–3 per 1000 live births, respectively.

FAS / FASD - definitions

FASD is an umbrella term describing the range of effects that can occur in an individual whose mother drank alcohol during pregnancy.

These effects may include physical, mental, behavioral, and learning disabilities with lifelong implications.

The term FASD is not intended for use as a clinical diagnosis.

FAS / FASD - definitions

The term **FAS** = health damage related mainly to central nervous system, dysmorphy and functional-behavioral features, in children who were prenatally exposed to ethanol.

1973 – first description and report of a disabled, delayed infant whose mother consumed alcohol during pregnancy.

Medical Dx FAS

4 fundamental criteria:

- Evidence of alcohol consumption during pregnancy
- Growth failure (pre- and post-natal), deficits in head size, microcephaly
- CNS damage, impaired intellectual, emotional and social functions
- Physical abnormalities – facial dysmorphia, defects of limbs and internal organs.

Table 1: Institute of Medicine diagnostic criteria for fetal alcohol syndrome and alcohol-related effects[4]

Fetal alcohol syndrome (FAS)

1. *FAS with confirmed maternal alcohol exposure**
   - A. Confirmed maternal alcohol exposure*
   - B. Evidence of a characteristic pattern of facial anomalies that includes features such as short palpebral fissures and abnormalities in the premaxillary zone (e.g., flat upper lip, flattened philtrum and flat midface)
   - C. Evidence of growth retardation, as in at least one of the following:
     - low birth weight for gestational age
     - decelerating weight over time not due to nutrition
     - disproportional low weight-to-height ratio
   - D. Evidence of central nervous system neurodevelopmental abnormalities, as in at least one of the following:
     - decreased cranial size at birth
     - structural brain abnormalities (e.g., microcephaly, partial or complete agenesis of the corpus callosum, cerebellar hypoplasia)
     - neurologic hard or soft signs (as age appropriate), such as impaired fine motor skills, neurosensory hearing loss, poor tandem gait, poor eye-hand coordination
2. *FAS without confirmed maternal alcohol exposure*

   B, C, and D as above
3. *Partial FAS with confirmed maternal alcohol exposure*
   - A. Confirmed maternal alcohol exposure*
   - B. Evidence of some components of the pattern of characteristic facial anomalies

   Either C or D or E
   - C. Evidence of growth retardation, as in at least one of the following:
     - low birth weight for gestational age
     - decelerating weight over time not due to nutrition
     - disproportionally low weight-to-height ratio
   - D. Evidence of CNS neurodevelopmental abnormalities, e.g.,
     - decreased cranial size at birth
     - structural brain abnormalities (e.g., microcephaly, partial or complete agenesis of the corpus callosum, cerebellar hypoplasia)
     - neurologic hard or soft signs (as age appropriate) such as impaired fine motor skills, neurosensory hearing loss, poor tandem gait, poor eye–hand coordination
   - E. Evidence of a complex pattern of behaviour or cognitive abnormalities that are inconsistent with developmental level and cannot be explained by familial background or environment alone: e.g., learning difficulties; deficits in school performance; poor impulse control; problems in social perception; deficits in higher level receptive and expressive language; poor capacity for abstraction or metacognition; specific deficits in mathematical skills; or problems in memory, attention or judgment.

If all 4 diagnostic criteria are present, including specific dysmorphic features
→ FAS (Fetal alcohol syndrome)

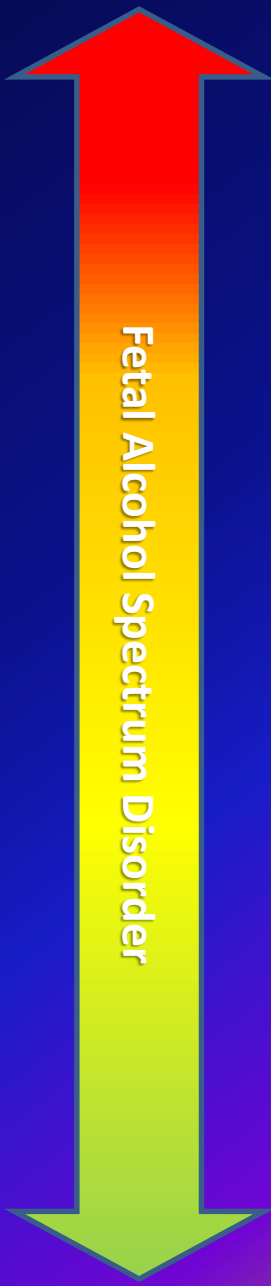
If less than 4 criteria:

FAE *(Fetal Alcohol Effect)*

ARND *(Alcohol-Related Neurodevelopmental Disorder)*

FARC *(Fetal Alcohol-Related Conditions)*

ARBD *(Alcohol-Related Birth Defects)*



FAS / FAE

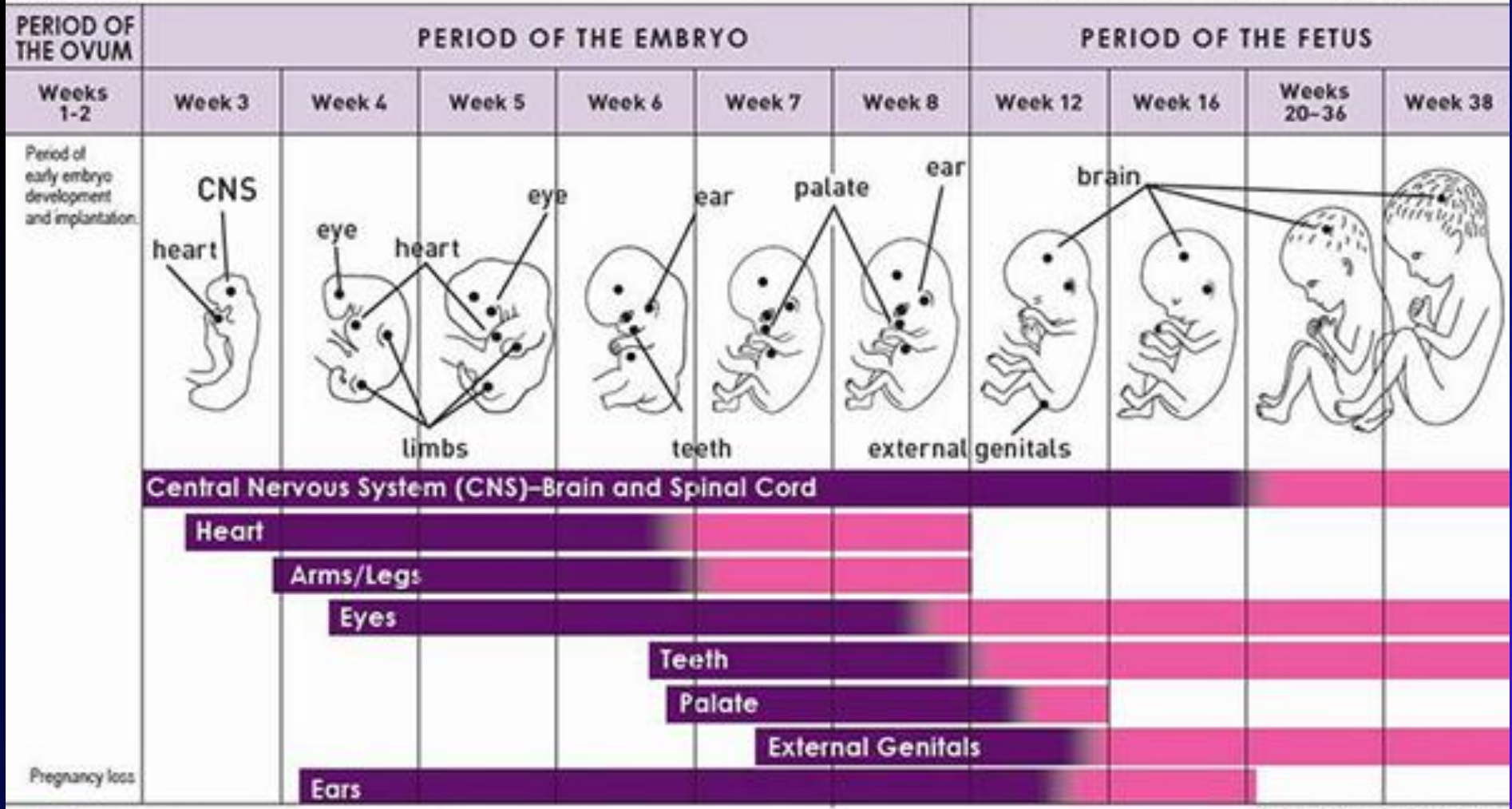
Incomplete features of FAS = ***Fetal Alcohol Effects*** **(FAE)**

Normal growth, appearance, mental status, however, behavioral problems, lower school performance and disturbed train of thought.

FETAL DEVELOPMENT CHART

This chart shows vulnerability of the fetus to defects throughout 38 weeks of pregnancy.*

• = Most common site of birth defects



Period of development when major defects in bodily structure can occur.

Period of development when major functional defects and minor structural defects can occur.

Adapted from Moore, 1993 and the National Organization on Fetal Alcohol Syndrome (NOFAS) 2009

*This fetal chart shows the 38 weeks of pregnancy. Since it is difficult to know exactly when conception occurs, health care providers calculate a woman's due date 40 weeks from the start of her last menstrual cycle.

Risk of alcohol use in pregnancy

I trimester:

- Brain damage
- Cellular intoxication
- Heart & kidney damage
- Facial malformations
- Abortion / Miscarriage

Risk of alcohol use in pregnancy

II trimester:

- Impaired brain development,
- miscarriages,
- Muscle, skin, glands, bones, and teeth damage.

III trimester:

- Impaired brain and lungs development,
- Growth failure.
- Withdrawn syndrome

Dysmorphism associated with FAS

Small head (microcephaly)

Small eye openings, downslanting of the eyes

Wide setting of the eyes (hypertelorism)

Epicanthal folds (epicanthus)

Short, rotated upwards nose, low nasal bridge

Smooth philtrum

Thin or lack of upper lip

Flat mid-face

Undeveloped jaw (micro- and retrognathia)

Low set ears

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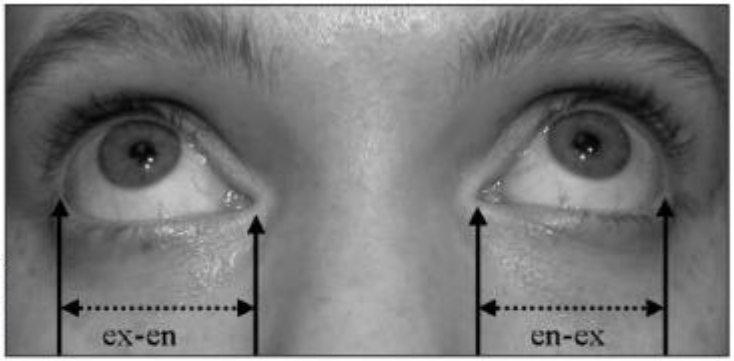

Fig. 2: Palpebral fissure length. To measure palpebral fissure length, identify the inner corner or encanthion (en) and outer corner or excanthion (ex) for each eye. Have the patient look up so that ex can be seen clearly. With a clear flexible ruler held in the horizontal plane, measure the length of each ex-en interval immediately below the eye, being careful not to touch the eye or eyelashes. Plot the result on an appropriate nomogram chart to determine the percentile or standard deviation for each eye.

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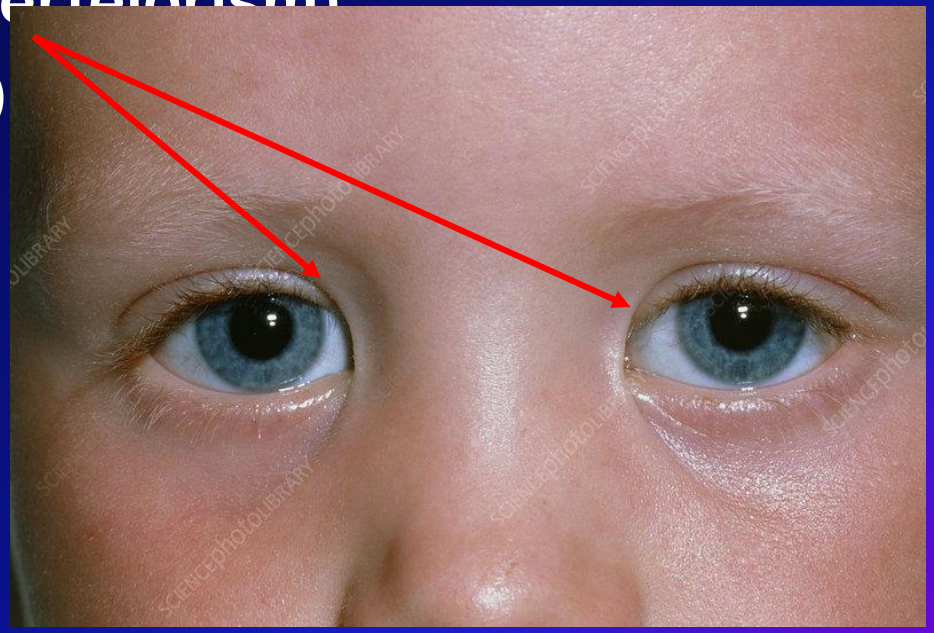

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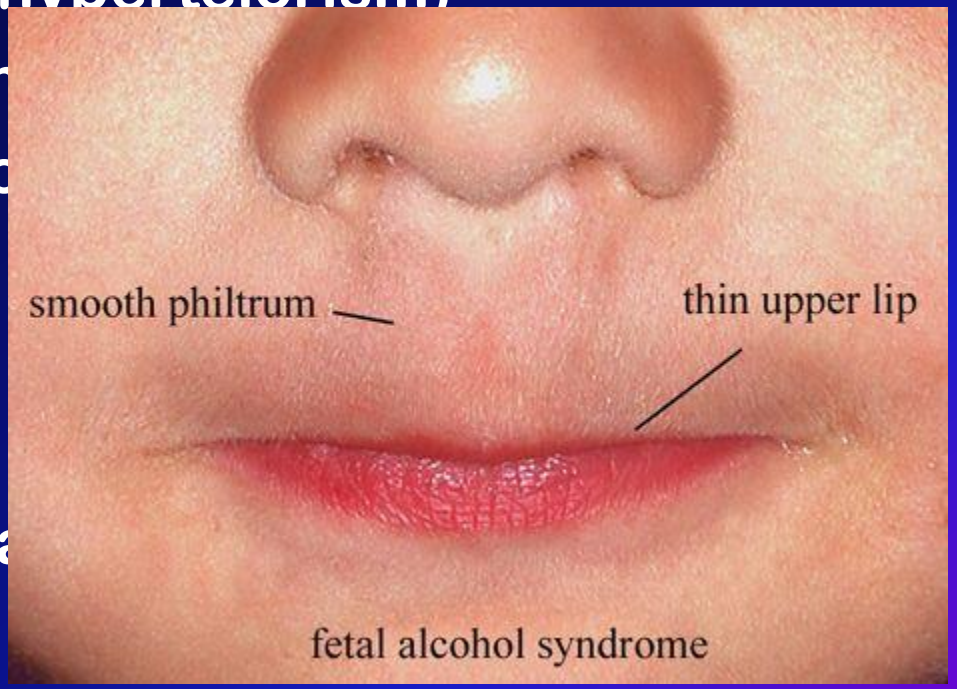
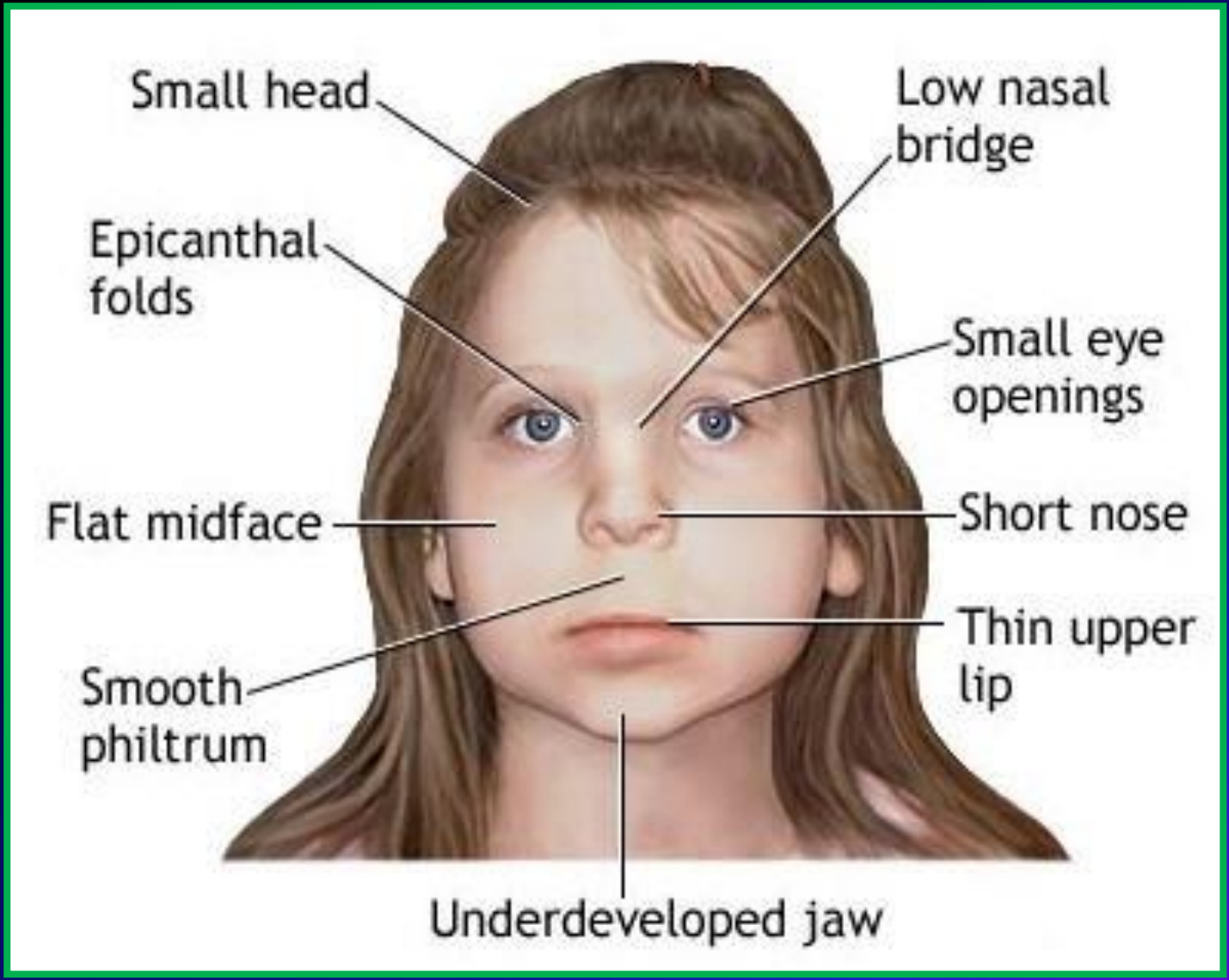
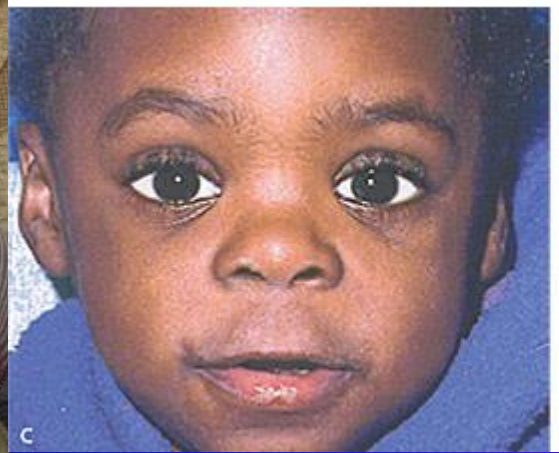


Dysmorphism in FAS




A


B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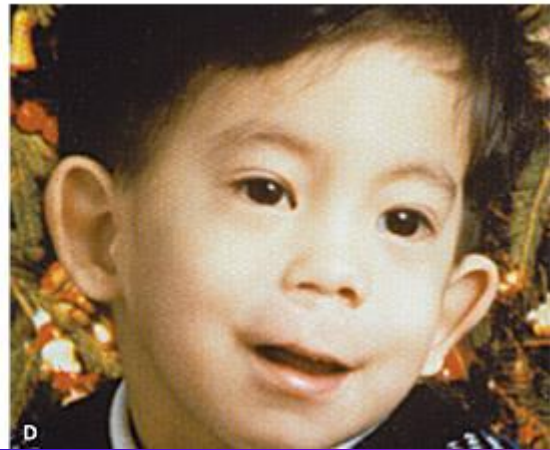
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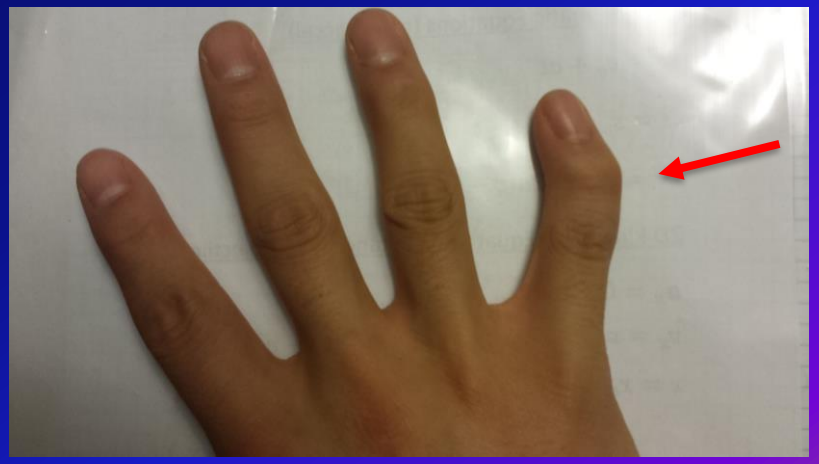
Congenital defects associated with FAS

Microcephaly	53 – 86%
• Heart	29 – 41%
• Genital deformities	46%
• Urogenital	10%
• Skeletal / Thoracic	27%,
• Hypoplasia of the hip	19%
• Clinodactyly of the 5th finger (hand)	22 – 51%



Cognitive function in FAS

Intelligence range – from 29 to 120 IQ,

Memory problems

Deficit in cause-efect thinking and analysis

Lack of the ability to generalize

Attention deficit

Hyperactivity

Difficulties in planing, predicting, imagination

Emotional signs in FAS

Atypical reacting to unknown or frustrating situations

Aggressive or withdrawn

Hyperactive…

Depressive / Frustrated

Lack of critical assessment of the situation and context

Impulsive

Submissive

Tendency to lie and steal / theft

For fetuses, there is no safe dose of alcohol to be consumed at pregnancy.

Even a minimal dose may severely disturb fetal development

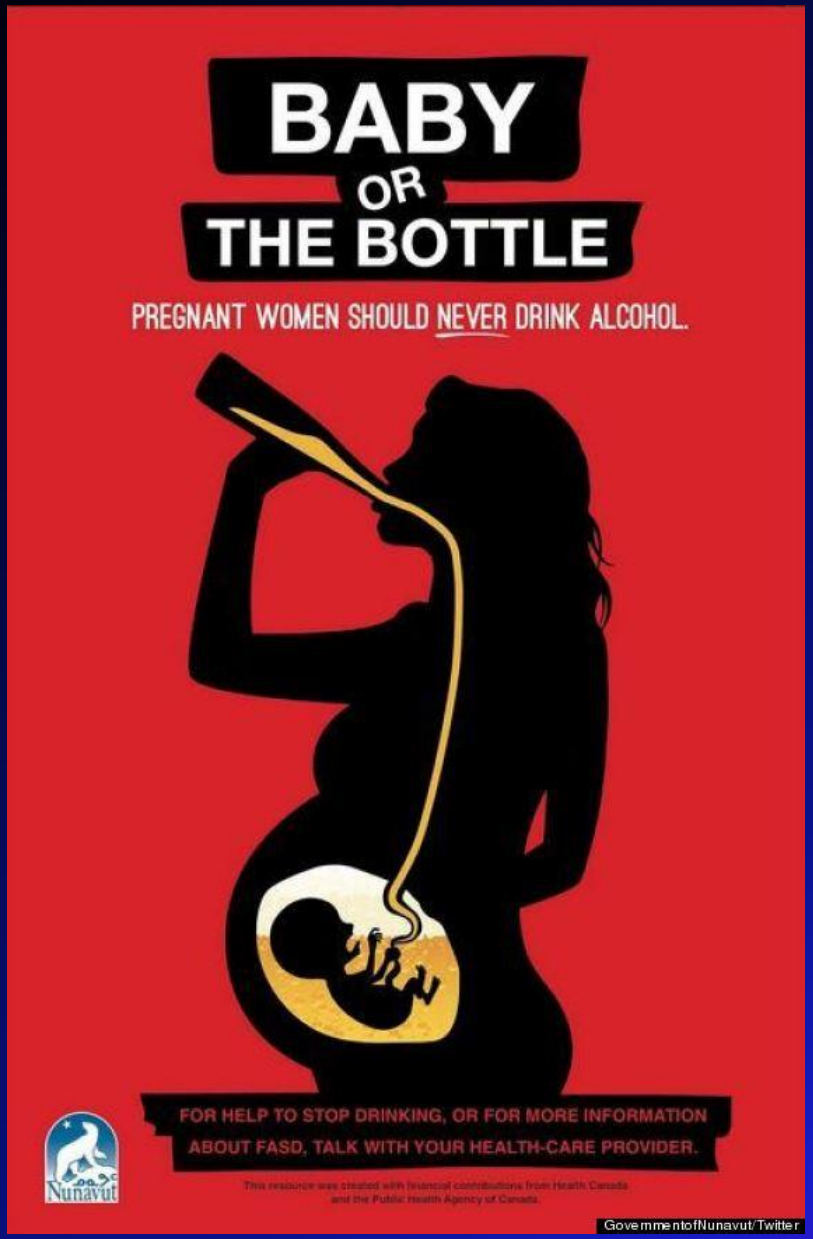
The consequences of alcohol consumption during pregnancy are at least as dangerous as intake of other drugs such as marijuana, heroinę, or cocaine.

FAS / FASD is an absolutely avoidable disease:

Just restrain from drinking alcohol while pregnant

Be aware

and beware


BABY
OR
THE BOTTLE
PREGNANT WOMEN SHOULD NEVER DRINK ALCOHOL.
FOR HELP TO STOP DRINKING, OR FOR MORE INFORMATION
ABOUT FASD, TALK WITH YOUR HEALTH-CARE PROVIDER.
Nunavut
GovernmentofNunavut/Twitter

Thank you