



# MICROCEPHALY

## DEFINITION



Defined as head circumference that measures 3 standard deviations below the mean for age and sex .

# MICROCEPHALY



# ETIOLOGY



- 2 Main Groups:
  - Primary (Genetic)
  - Secondary (Non Genetic)

# PRIMARY MICROCEPHALY



A group of conditions that usually have no associated malformations and follow a mendelian pattern of inheritance or are associated with specific genetic syndrome.



## CAUSES OF MICROCEPHALY

- **FAMILIAL**
  1. Autosomal dominant
  2. Autosomal recessive
  
- **SYNDROMES**
  1. Downs syndrome
  2. Edward syndrome
  3. Cri-du-chat
  4. Cornelia de lange
  5. Rubinstein - taybi

## SECONDARY MICROCEPHALY



Results from insult that affect in-utero or an infant during periods of rapid brain growth especially during 1<sup>st</sup> year of life.

# CAUSES OF MICROCEPHALY- CONTD..



## CONGENITAL INFECTIONS

- Cytomegalovirus
- Rubella
- Toxoplasmosis

## DRUGS

- Fetal Alcohol
- Fetal Hydantoin

# CAUSES OF MICROCEPHALY- CONTD..



## MISCELLANEOUS

- Metabolic (maternal diabetes, maternal, hyperphenylalaninemia)
- Meningitis
- Hyperthermia
- Hypoxic –ischemic encephalopathy
- Radiation

# EVALUATION - HISTORY



## Antenatal

- Fever with rash
- Maternal hyperthermia
- Radiation exposure
- Drug intake - phenytoin

# EVALUATION - HISTORY



## Postnatal

- Birth asphyxia
- CNS infections
- Head circumference at birth

## Family history

- Small head
- Mental retardation
- Seizures

# EXAMINATION



- Head circumference(occipitofrontal circumference - OFC)
- To record the greatest volume of cranium.

## MEASUREMENT OF OFC



- A fibroelastic tape is placed over the nasion anteriorly and most prominent portion of occiputoposteriorly
- Measurement made by over lapping tape over temporal bone.

# MEASUREMENT OF OFC



# INCREMENT OF HEAD CIRCUMFERENCE



- Normal value at birth 34-35 cm

| AGE        | GROWTH IN HEAD<br>CIRCUMFERENCE(cm/month) |
|------------|-------------------------------------------|
| 0-3 month  | 2                                         |
| 3-6 month  | 1                                         |
| 6-9 month  | 0.5                                       |
| 9-12 month | 0.5                                       |
| 1-3 yrs    | 0.25                                      |
| 4-6 yrs    | 1cm/yr                                    |

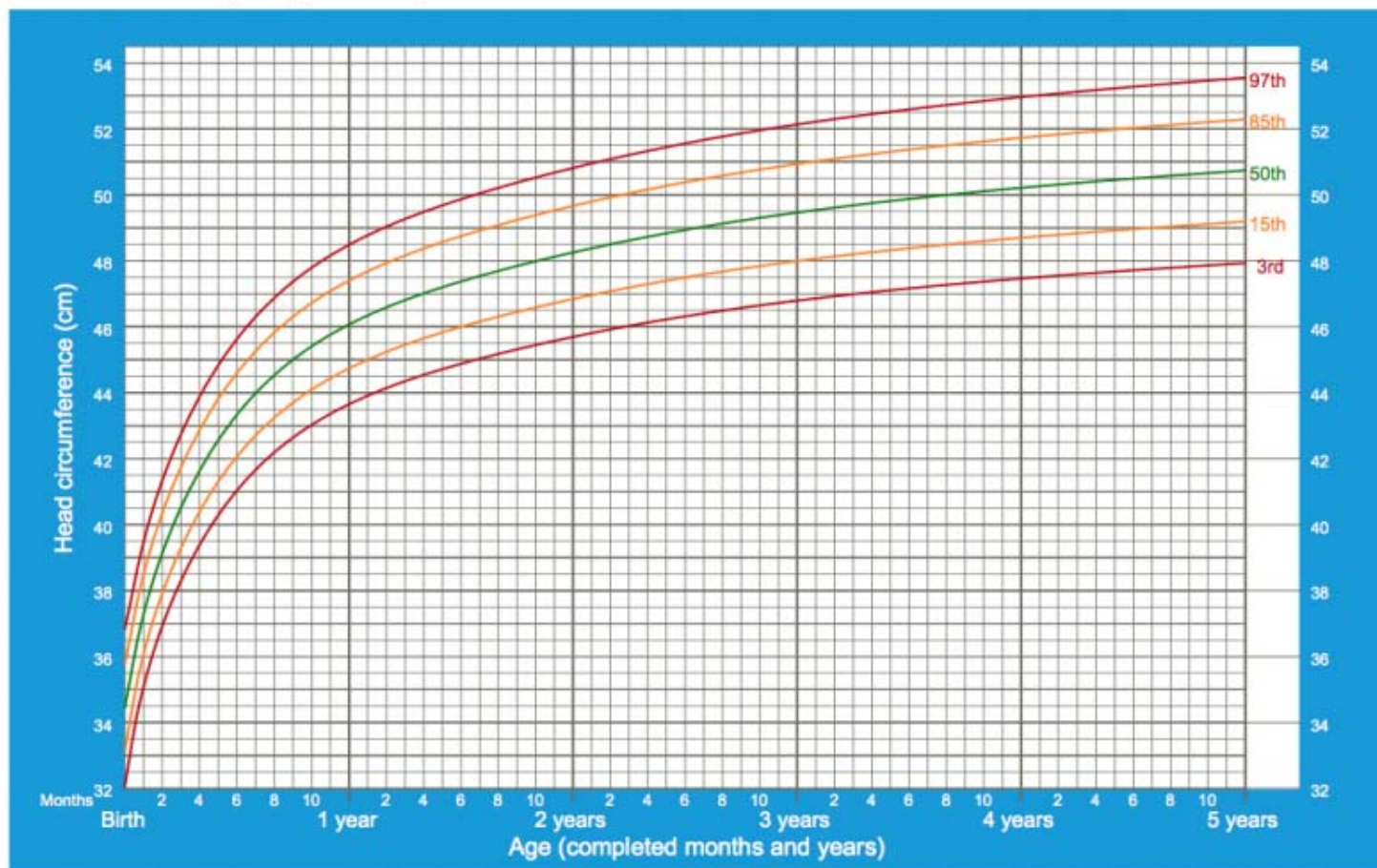
# HEAD CIRCUMFERENCE



- Head circumference can be plotted on growth charts
- Serial measurements are more meaningful
- Head circumference of parents and siblings should be recorded

## Head circumference-for-age BOYS

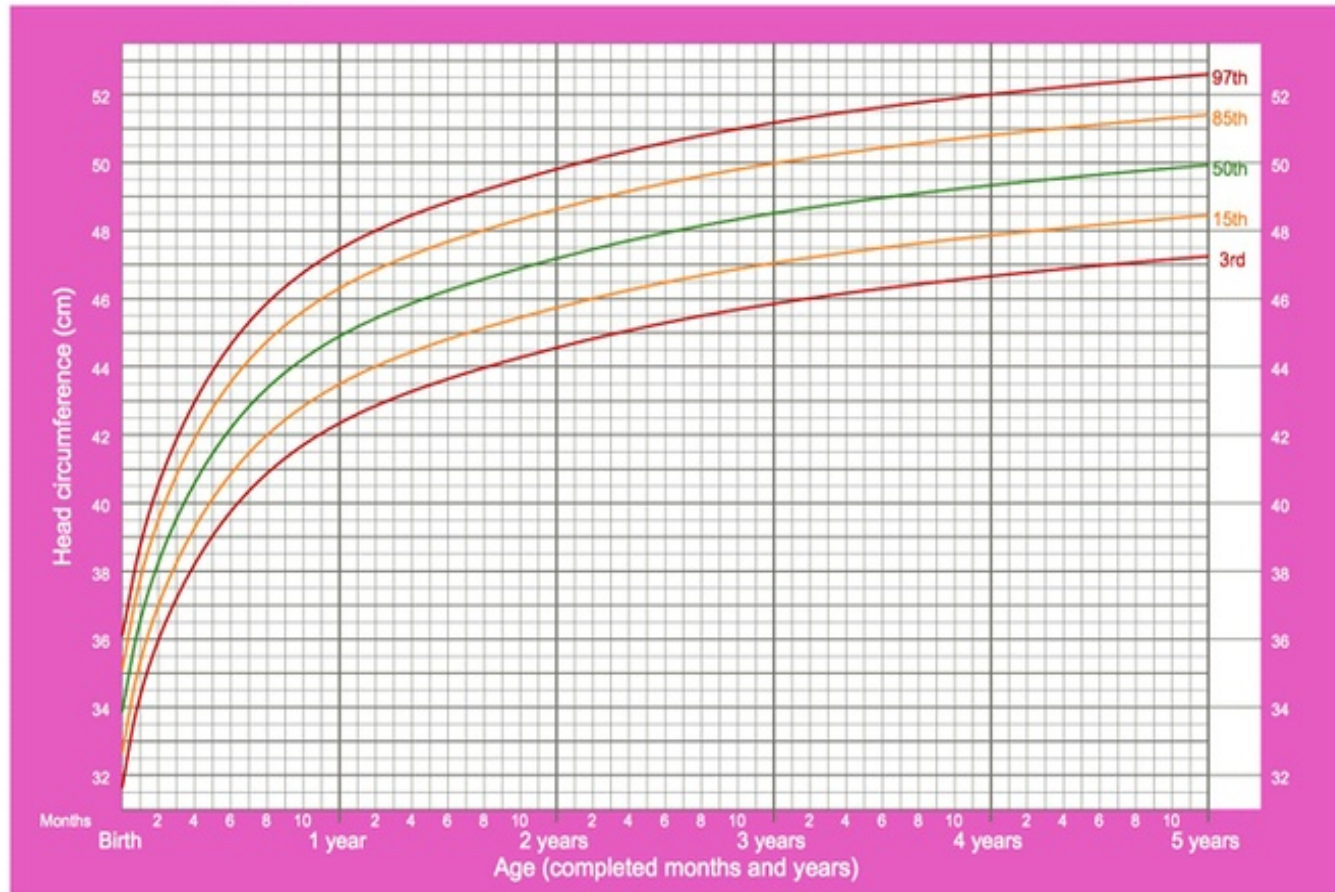
Birth to 5 years (percentiles)



WHO Child Growth Standards

## Head circumference-for-age GIRLS

Birth to 5 years (percentiles)



WHO Child Growth Standards



## DYNE'S FORMULA

- Head circumference length ratio(Dyne et al) applied to infants below 1 yr of age
- Normal HC in cms =  $\text{length in cm}/2 + (9.5 \pm 2.5)$

## EXAMINATION-CONTD



- Abnormal head shape
- Fontanel
- Sutures
- Stigmata of intrauterine infections
- Dysmorphism
- Detailed CNS examination



## INVESTIGATION

- Determined by history and physical examination
- TORCH titers-intrauterine infections
- Karyotyping-chromosomal syndrome
- Maternal serum phenylalanine - (PKU)

## INVESTIGATION-CONTD



- Fetal USG-if family history of small head
- CT scan-intracerebral calcifications
- MRI scan-structural abnormality of brain



## TREATMENT

- Head size cannot be changed with treatment
- Genetic & family counseling provided
- Associated problems –
  - i. Hearing/Visual Problems
  - ii. Mental Retardation
  - iii. Seizures

# THANK YOU

