MACROCRANIA

True Megalencephaly

Exclusion of Secondary Causes of Macrocrania

Anatomic/developmental Megalencephaly
- Symmetric or asymmetric
- Usually stable imaging findings
- Other cortical and midline malformations are common

Assess extent and location of the parenchymal enlargement
- Involvement of the 2 cerebral hemispheres +/- posterior fossa structures
- Enlargement 1 whole cerebral hemisphere
- Enlargement 1 whole cerebral hemisphere + ipsilateral brainstem and cerebellum
- Enlargement of part of 1 cerebral hemisphere A/P gradients

Bilateral MEG
Unilateral MEG/ HMEG
“Total” HMEG
“Quadrantic dysplasia”/“Lobar HMEG”/ “Hemi-HMEG”

Assess presence of any of the following:
- Cortical malformations
- White-matter abnormalities
- CC and other midline anomalies
- Acquired tonsillar ectopia

Metabolic Megalencephaly
- Usually symmetric
- Frequent imaging changes over time
- Other intracranial malformations usually absent
- Supporting imaging features: bilateral signal changes in GM/WM, parenchymal cysts, areas of restricted diffusion or contrast enhancement
- For specific entities: pattern recognition + clinical features!

H-MRS: may add important info (eg, increased NAA peak in Canavan disease)