Infantile hemangiomas

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Infantile Hemangioma (IH)

<table>
<thead>
<tr>
<th>Risk factors</th>
<th>Pathogenesis</th>
<th>Clinical</th>
<th>Pathology</th>
<th>Complications</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Caucasian</td>
<td>Not fully elucidated. Theories include: Vasculogenesis &amp; angiogenesis</td>
<td>Clinical types</td>
<td>Proliferative phase</td>
<td>Uleration</td>
<td>Topical</td>
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<td>2. Female</td>
<td>- 1 VEGF signalling → endothelial cell proliferation</td>
<td></td>
<td>Lobular endothelial proliferation</td>
<td>- Most common complication, up to 10%</td>
<td>- Timolol 0.5% (max 0.25mg/kg/day)</td>
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<td>3. Higher maternal age</td>
<td>- Expression of placenta-associated vascular antigens (GLUT-1)</td>
<td></td>
<td>Involution phase</td>
<td>- IH at risk: on lips, anogenital, skin folds, large, mixed, or segmental IH</td>
<td>- Superpotent corticosteroids</td>
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<td>4. Prematurity</td>
<td>- Hypoxia</td>
<td></td>
<td>Fibrous &amp; fatty tissue</td>
<td>- Increased risk of infection &amp; scarring</td>
<td>Intralerial</td>
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<td>5. Low birth weight</td>
<td>- 1 GLUT1 &amp; VEGF mobilization of endothelial progenitor cells</td>
<td></td>
<td></td>
<td>Disfigurement, functional impairment</td>
<td>- Tramcinolone 5-40mg/ml (max 3-5mg/kg)</td>
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<tr>
<td>6. Multiple gestation</td>
<td>- Genetic associations: VEGFR2, ANTXR1, loss of heterozygosity of Sq</td>
<td>Patterns of involvement</td>
<td></td>
<td></td>
<td>Systemic</td>
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<td>7. Placental insufficiency</td>
<td>- Focal</td>
<td></td>
<td>- Periocular, nasal tip, columella, lip, pinna, breast, anogenital IH</td>
<td></td>
<td>- Indications for systemic therapy: lesions threatening vision/airway, liver involvement (or high output CHF), risk for disfigurement, ulceration</td>
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<td>8. Chorionic villus sampling</td>
<td>- Multifocal: if ≥5 lesions are present</td>
<td></td>
<td>Extracutaneous involvement</td>
<td></td>
<td>- Propranolol (1st line)</td>
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</tbody>
</table>

Syndromes associated with segmental hemangiomas

- Posterior fossa & other brain malformations: Dandy-Walker, cerebellar hypoplasia
- Hemangiomas: segmental (face & neck)
- Arterial abnormalities: cervical & cerebral artery aplasia, dysplasia, aneurysms (*terebrovascular anomalies = most common)
- Cardiac defects: aortic arch abnormalities, VSD, ASD
- Eye abnormalities: retinal vascular abnormalities, optic nerve hypoplasia
- Sternal defects & supraventricular raphae
- Lumbosacral/Lower body hemangioma & Lipomas or other cutaneous anomalies (“skin tags”)
- Urogenital anomalies
- Myelopathy (spina bifida)
- Bony deformities (hip dysplasia, leg length/width discrepancy, scoliosis)
- Anorectal (fistula, imperforate anus) & Arterial anomalies (lower limb stenosis, dysplasia)
- Renal anomalies (hypoplastic, single, pelvic kidney)

Congenital hemangiomas and hallmark features that differentiate them from infantile hemangiomas

- Fully formed at birth
- Pathophysiology: Most have mutation in GNAQ or GNA11
- Doppler: dense vascularity, fast-flow
- Rapidly involuting congenital hemangiomas (RICH): rapidly involute in first year of life
- Non-involuting congenital hemangiomas (NICH): do not involute, grow proportionally with child, may be painful
- Partially involving congenital hemangiomas (PICH): intermediate form, undergoes partial involution
- Pathology: Negative GLUT1 and Lewis Y antigen


References