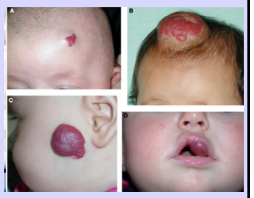
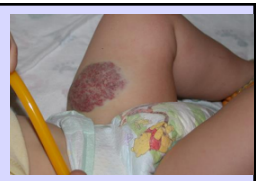
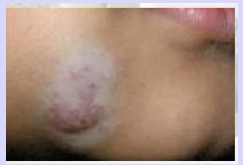
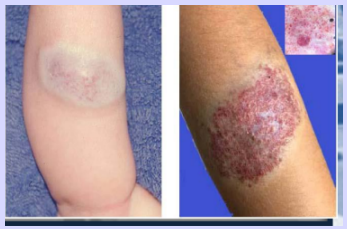
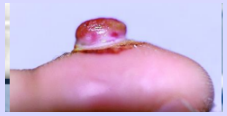
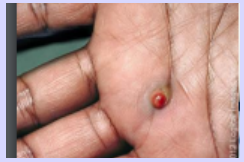
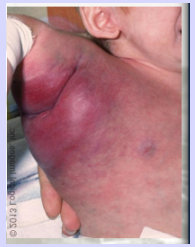
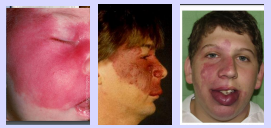
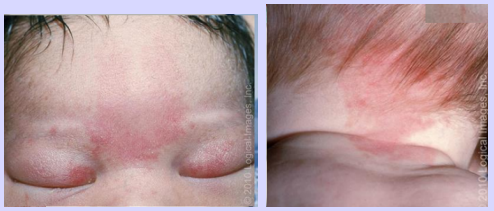
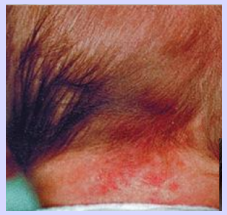
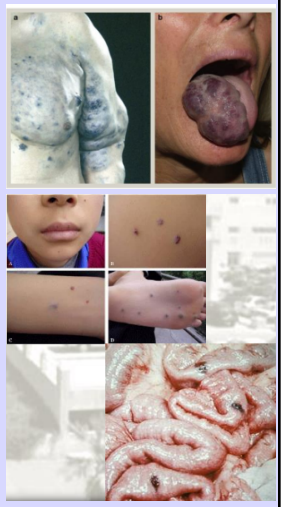
Vascular Anomalies:

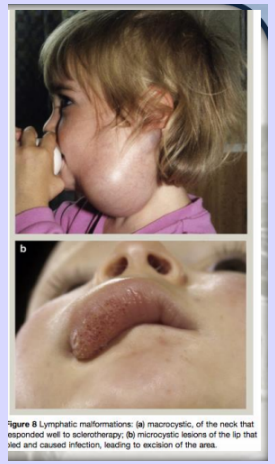
1. **Vascular tumors**:
    a. Benign:
        I. Infantile hemangioma
        II. Congenital hemangioma:
            1. Rapidly Involuting Congenital hemangioma (RICH)
            2. Non-Involuting Congenital hemangioma (NICH)
            3. Partially Involuting Congenital hemangioma (PICH)
        III. Tufted angioma
    b. Borderline / locally aggressive:
        I. Kaposiform
        II. Hemangioendothelioma
        III. Kaposi Sarcoma
    c. Malignant
        I. Angiosarcoma
    d. Associated with other lesions:
    Posterior fossa malformations
    Hemangioma
    Arterial anomalies
    Cardiovascular anomalies
    Eye anomalies
    Sternal cleft/ supraumbilical raphe

2. **Vascular anomalies**
    a. Simple:
        I. Slow flow:
            1. Capillary malformations (CM)
            2. Venous malformations (VM)
            3. Lymphatic malformations (LM)
        II. High flow:
            1. Arteriovenous malformations (AVM)
            2. Arteriovenous fistula (AVF)
    b. Combined:
        I. CVM
        II. CLM
        III. LVM
        IV. CAVM

Benign Vascular Tumors	**Presentation**	**Features**	**Management / Treatment**	**Picture**
Infantile Hemangioma (Strawberry naevi)	head /neck Stage 1 (5-8 m): Rapid disfiguring growth, soft and warm, with a prominent Doppler signal. Stage 2 (7-9 y): prolonged, darkening with grey hue, slow loss of color, fine capillary telangiectasia Stage 3 (7-9 y): visible soft lump regression, cellular parenchyma replaced with fibro-fatty residue	Localized / diffused Expression of GLUT-1 protein histological resemblance to placental tissue PHACE association	Expectant, Rarely biopsy CBC MRI/ US active intervention: – large size/ disfigurement – multiple lesions causing high-output cardiac failure – obstruction of vital structures (vision, airway) – persistent ulceration. 1st line: Propranolol: Cause vasoconstriction 1-2mg/kg/d 2nd line: steroids: Intralesional 2mg/kg (4-6 w) Systemic therapy: Rebound growth Embolization: in high output cardiac failure / bleeding lesions surgical excision, Tracheostomy Pulse-dye laser: in surface residual telangiectasia (>10 y) coagulate surface of ulcerated lesions (dressing is 1ry care)	
RICH	Large mass on leg Faster involution, full regression by 1 year of age	negative for GLUT-1 firmer than infantile haemangiomas, with or without telangiectatic changes Leave plaque-like residuum, may regress leaving atrophic patch of skin		

NICH	mimic infantile haemangiomas and of similar texture	round /oval masses, with flat shape or moderately bossed and accompanying telangiectasia, and may have a halo. do not exhibit further growth and do not regress	Expectant management surgical excision	 
PICH	variant which looks like a NICH but slowly regresses by age 10	-		
Pyogenic granuloma (PG) Lobular Capillary Hemangioma	starts as small red papule that grows rapidly over weeks to months and then stabilizes	Rapid growth, friable surface: bleeds profusely after minor trauma and may become ulcerated. Bleeding difficult to control recurrent		
Tufted Angioma	Round or oval masses, flat shape or moderately bossed, telangiectasia, no growth or regression	-	Expectant management Observation, may require surgical excision	
Kaposiform (locally aggressive)	Kasabach-Merritt phenomenon KMP		MTOR +ve : Sirolimus	

Vascular malformations	presentation	features	management	picture
CM	Port wine stain (newborns)	colour deformity may cause psychological concern and impair normal social interaction. – In teenager and adults tissue hypertrophy may cause further concern	supportive with involvement of a clinical psychologist, with camouflage and the use of pulse dye laser therapy. – Which can lighten the colour for a number of years. Surgery may be useful for reducing hypertrophied areas: the lower lip	
Nevus simplex (macular stain, salmon patch, stork bite, angel kiss)	single or multiple blanchable, pink-red patches in newborn infants (infants) most commonly on eyelid, glabella, and midline of nape of neck. Less common sites: scalp, nose, lip, and back	generally fades within one to two years, although lesions on the back of the neck may persist unchanged with little consequence		
VM	Disfigurement Pain Coagulopathy: D-dimer/ fibrinogen	low-flow lesions are blue, compressible soft tissue masses that empty on elevation. They can affect most tissues 5% genetic abnormalities – Krit-1, TIE-2 and Glomulin genes – Blue rubber bleb syndrome	Compression garments NSAIDS Sclerotherapy Surgery	
Combined Lesions	significant morbidity with painful, heavy areas involving a limb,	isolated or associated with overgrowth disorders such as Klippel- Trenaunay and Proteus syndrome	require life-long care	

	episodes of infection, wound breakdown			
LM	-	Microcystic / macrocystic	Sclerotherapy – OK-432 Surgery – Seroma – Infection	
AVM	Schobinger classification: Stage 1 (Quiescence): pink/ blue stain, warmth, AV shunt Stage 2 (Expansion): Stage 1 + enlargement, pulsations, thrills, bruits Stage 3 (Destruction): Stage 2 + dystrophic skin changes, ulceration, bleeding, pain / tissue necrosis Stage 4 (Decomposition): Stage 3 + high output cardiac failure	High-Flow Lesions, Characteristic nidus with arterial feeders, arteriovenous fistulas, enlarged veins	symptomatic stages (III and IV): Interventional radiology, surgical excision, reconstruction, repeated embolization Embolic agents include • ethanol, cyanoacrylate (glue), coils, polyvinyl particles and onyx, a liquid ethylene vinyl alcohol copolymer	

Molecular mechanism:

The mammalian target of rapamin (MTOR) pathway22 is an intra- cellular signalling pathway which results in cell growth and survival

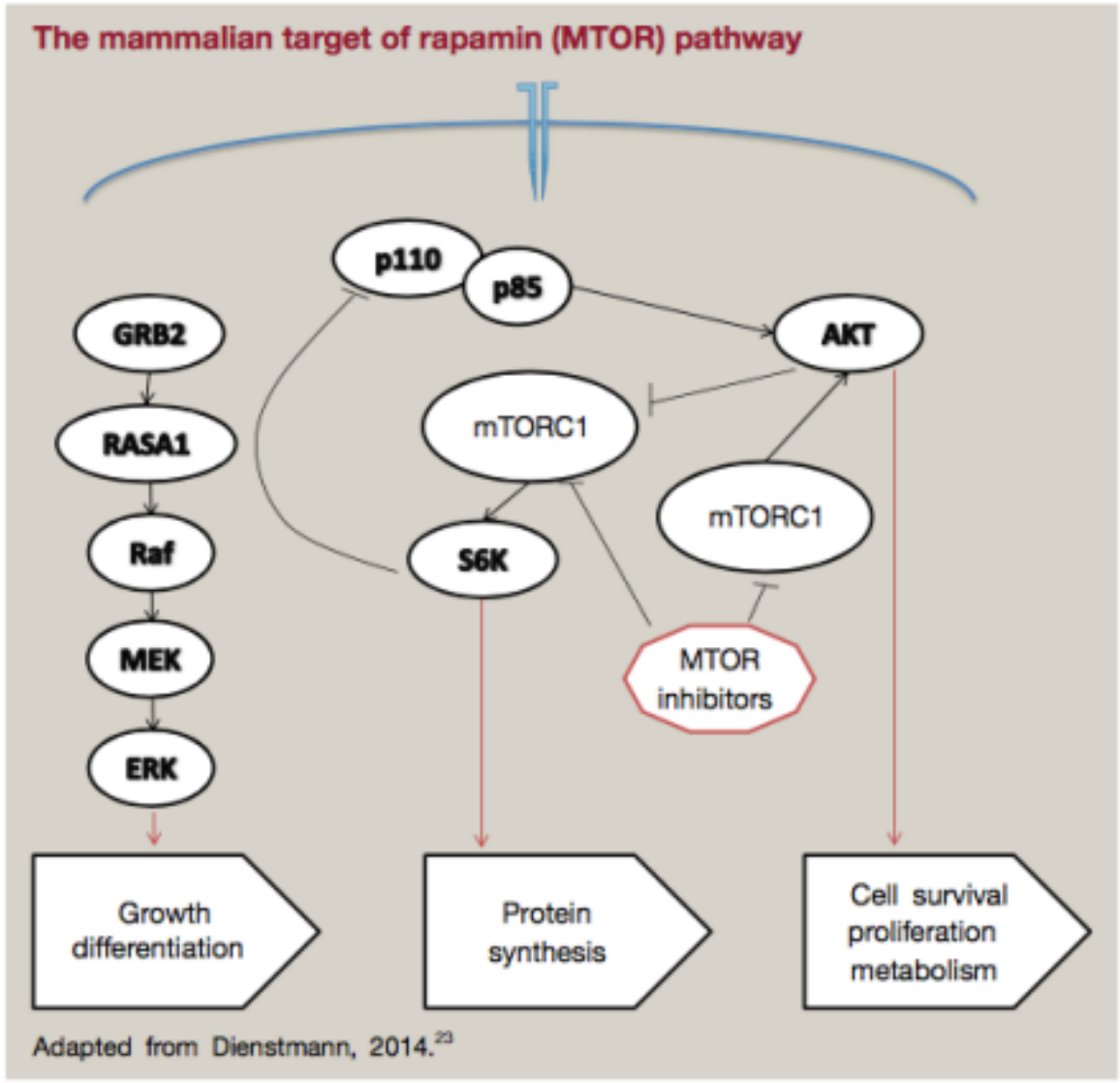


Figure 10 The mammalian target of rapamin (MTOR) pathway Adapted from Dienstmann, 2014.[22]