Background:
Vascular anomalies comprise a heterogeneous group of disorders. The presence of D2-40 markers and the Kasabach-Merrit phenomenon (KMP), are associated with a major gravity.

Objectives:
To analyze the efficacy and safety of treatment with sirolimus in children with complicated vascular anomalies (CVA).

Results:

**CASE 1**
14-month-old male
Lymphangiomatosis in his right upper extremity
Rehabilitation treatment failure

Sirolimus 0.8 mg/m²/12h
Plasma level peak: 13.22 ng/mL [6.27- 26.19]

Dose adjustment: 0.4 mg/m²/day (azithromycin concomitance)

After 262 days with active treatment, objective clinical improvement in the functionality of the affected limb was achieved.

No adverse effects were observed

**CASE 2**
32-month-old male
Unresectable cervical kaposiform hemangioendothelioma (KMP) treated with acetylsalicylic acid + ticlopidine, previously treated with vincristine and systemic high-dose glucocorticoids

Sirolimus 0.8 mg/m²/12h
Plasma level peak: 9.86 ng/mL [3.49- 17.8]

Adverse effect: Hypertriglyceridemia

Dose reduction: 0.8 mg/m²/day
Plasma level peak: 3.73 ng/mL [2.9- 4.95]

Platelet values at fifth day and maintained normal during all the treatment (388 days), and 88 days after stopping it.

Conclusions:
Sirolimus has been shown as an effective therapeutic option for CVA in childhood. It was well tolerated, and adjusting plasma levels allowed adverse effects minimisation without compromising effectiveness. Further studies are needed to determine the contribution of mTOR inhibitors in the treatment of childhood vascular anomalies.