



Kasabach-Merritt phenomenon (KMP)

This information sheet provides information about Kasabach-Merritt syndrome, what causes it and how it can be treated. It also explains what to expect when your child comes to Great Ormond Street Hospital (GOSH) for diagnosis and treatment.

What is Kasabach-Merritt phenomenon (KMP) and what causes it?

Kasabach-Merritt phenomenon (KMP) refers to clotting problems arising as a result of the rare benign (noncancerous) vascular lesions known as kaposiform haemangioendothelioma (KHE) and tufted angioma. Information about these conditions is available on our website. Infantile haemangiomas never lead to KMP.

KMP arises because the KHE or tufted angioma causes trapping of platelets. As the lesion grows, more platelets are trapped, causing increasing impairment of clotting.

How common is KMP?

KMP is very rare affecting only a few children with very rare types of vascular lesion.

What are the symptoms of KMP?

The main symptoms associated with KMP are due to a tendency to bleed that arises from lack of platelets. The medical term for this is thrombocytopenia. If a child has too few platelets circulating in the blood, they bruise more easily than usual or develop tiny pinprick bruises under the skin called petechiae.

How is it diagnosed?

KMP is usually suspected when a child presents with a specific type of vascular lesion. It is confirmed by taking blood samples for analysis in the laboratory. In most cases, the lesion will have been diagnosed before the bruising or bleeding starts.

Can KMP be treated?

KMP can be corrected. As the severity of symptoms varies from child to child, a multidisciplinary team in a specialist setting often provides the best care. Dermatologists (doctors specialising in skin disorders) and haematologists (doctors specialising in blood disorders) are usually involved.

If a child has only a slightly low platelet count and mild clotting problems, they might not need any active treatment but will need regular monitoring to ensure that problems are identified at an early stage.

If a child has more severe platelet and clotting problems, treatment may be with vincristine or rapamycin (sirolimus) – these medications will be discussed further with you. Embolisation by an interventional radiologist may be suggested to block the blood vessels in the lesion, with the aim of stopping it growing and improving the platelet count. Very occasionally, depending on the size and location of the lesion, surgery might be appropriate. In the most serious cases, children may need to have regular blood transfusions.

What is the outlook for children with KMP?

KMP can be a very serious disease. Treatment is intensive, but is successful in most cases.

Information and support

At Great Ormond Street Hospital (GOSH), contact the Birthmark Unit.

The Birthmark Support Group offers support and advice to parents of children with all types of birthmark, including KMP. Telephone their helpline on 0845 045 4700 or visit their website at www.birthmarksupportgroup.org.uk.