Klippel-Trenaunay Syndrome

Frequently Asked Questions

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Frequently Asked Questions About KT-Syndrome

1. What is KT- Syndrome?

Described in 1900 by two French physicians; Klippel and Trenaunay, KT-syndrome is a rare combined and complex syndrome of vascular, bone and soft tissues with characteristic clinical features.

2. What are the main clinical presentations of Klippel-Trenaunay Syndrome?

The Salient features of KTS include a triad of varicose veins predominantly affecting the lower extremity, bone and soft tissue hypertrophy (enlargement), usually unilateral (affecting one side), as well as an overlying capillary malformation (Port wine stain).

3. What causes KT- Syndrome?

Although clinical research is ongoing and our insight into KT syndrome continues to expand KTS is thought to be a sporadic congenital phenomenon.

4. How does KTS affect individual patients?

The degree of involvement of KTS can be considerably variable in different people. Some patients can have a very mild presentation while others may have significantly complicated clinical findings. There may be small limb length discrepancy in some patients while others may have large and longer limbs with limitation of routine daily activities.

5. When does KTS appear?

Features of KTS are generally present at birth. However in some cases the signs may not be initially apparent.

6. What are the common symptoms of KT-Syndrome?

Depending on the degree and type of involvement of the KTS there can be different clinical scenarios. However patients generally complain of pain, fatigue and swelling of the involved limb, difficulty with ambulation, frequent cellulitis (infection of the skin), bleeding from varicose veins, or visceral bleeding such as rectal , or genitourinary bleeding. There can also be thrombosis of the deep veins of the lower extremities as well as pulmonary embolism (clots in the lungs).

7. How is KTS diagnosed?

The diagnosis of KTS is generally speaking on clinical grounds. However the importance of other diagnostic studies in patients with KTS cannot be refuted. These include MRI/MRA, venous ultrasonography, lymphscintigraphy among other modalities.

8. What are the treatment approaches for KT- Syndrome?

Treatment of KTS is generally that of a conservative medical regimen. It is geared toward prevention and treatment of skin infection with topical ,oral and parenteral antibiotics, reduction of swelling of the affected limb with compression stockings.

Other modalities include surgical debulking of the soft tissue which is generally suboptimal in terms of efficacy, treatment of varicose veins and limb length altering orthopedic procedures which can be an arduous undertaking fraught with potential complications.

9. How should a patient with KTS be evaluated?

Given the rare and complex nature of KT-syndrome with its variable degree of clinical symptoms it is felt that the patient will likely benefit from evaluation at a facility with familiarity with these types of vascular disorders.

Management of patients with KTS generally requires a multidisciplinary (team) approach as it is virtually impossible for a single clinician to evaluate or treat all aspects of KT-syndrome.

10. Is there a clinic at Minnesota Vascular and Birthmark Center (MV&BC) for KT-syndrome?

***Minnesota Vascular and Birthmark Center** is recognized internationally for its expertise in the diagnosis and management of patients with KT- syndrome. The Center has a dedicated referral clinic for patients with Klippel-Trenaunay Syndrome.*

To date patients from 32 states from around the United States have come to MV&BC for evaluation and treatment.

Dr. Delfanian, the medical director of Minnesota Vascular and Birthmark Center is a leading expert in the field of vascular anomalies and has special skills with respect to Klippel-Trenaunay syndrome.

He is a volunteer consultant for vascular Birthmark foundation providing clinical and educational services to patients, their families as well as communicating and collaborating with physician colleagues who may encounter a patient with KT- syndrome.