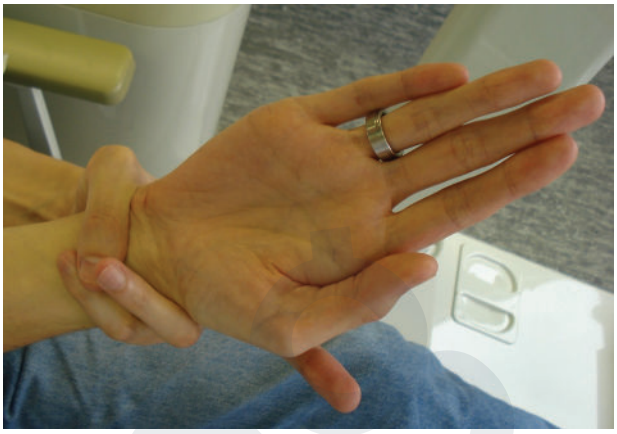
MARFAN SYNDROME PATHOCHART

PATHOPHYSIOLOGY

Marfan syndrome is a genetic condition in which the proteins that make up the connective tissue that supports the heart, blood vessels and other vital organs is weakened. Most people with this condition have heart and blood vessel problems, often resulting in aortic aneurysm or valve dysfunction. Patients with Marfan syndrome have tall, thin frames, long legs, fingers and hands and may have problems with vision.



By Staufenbiel I, Hauschild C, Kahl-Nieke B, Vahle-Hinz E, von Kodolitsch Y, Berner M, Bauss O, Geurtsen W, Rahman A - https://www.ncbi.nlm.nih.gov/pubmed/24165013, CC BY-SA 2.0, https://commons.wikimedia.org/w/index.php?curid=42107027

ASSESSMENT FINDINGS

- Tall and thin body type
- Flexible and/or painful joints
- Retracted or protruding sternum
- Disproportionately long arms, legs and fingers
- Scoliosis
- Heart murmur
- Vision difficulties

DIAGNOSTICS

- Cardinal features (at least 1)
  - Aortic root aneurysm
  - Dislocated lens of the eye
- FBN1 gene mutation
- Family history
- Ghent criteria

NURSING PRIORITIES

- Promote comfort
- Ensure adequate perfusion
- Optimize mobility and functional ability

THERAPEUTIC MANAGEMENT

- Symptom management
- Surgical correction of physical deformities or heart valve disorders
- Range of motion and weight-bearing exercises
- Treatment aimed at reducing cardiovascular compromise

MEDICATION THERAPY

- Beta-Blockers
- Calcium Channel Blockers
- Angiotensin Receptor Blockers
- Analgesics

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