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REVIEW ARTICLE

Marfan Syndrome: new diagnostic criteria, same anesthesia care? Case report and review

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Abstract
Background: Marfan's Syndrome (MFS) is a disorder of connective tissue, mainly involving the cardiovascular, musculoskeletal, and ocular systems. The most severe problems include aortic root dilatation and dissection. Anesthetic management is vital for the improvement on perioperative morbidity.
Case report: 61-year-old male with MFS, presenting mainly with pectus carinatum, scoliosis, ectopia lens, previous spontaneous pneumothorax and aortal aneurysm and dissection submitted to thoracoabdominal aortic prosthesis placement. Underwent routine laparoscopic cholecystectomy due to lithiasis. Important findings on preoperative examination were thoracolumbar kyphoscoliosis, metallic murmur on cardiac exam. Chest radiograph revealed Cobb angle of 70°. Echocardiogram showed evidence of aortic mechanical prosthesis with no deficits.
Discussion: Preoperative evaluation should focus on cardiopulmonary abnormalities. The anesthesiologist should be prepared for a potentially difficult intubation. Proper positioning and limb support prior to induction is crucial in order to avoid joint injuries. Consider antibiotic prophylaxis for subacute bacterial endocarditis. The patient should be carefully positioned to avoid joint injuries. Intraoperatively cardiovascular monitoring is mandatory: avoid maneuvers that can lead to tachycardia or hypertension, control airway pressure to prevent pneumothorax and maintain an adequate volemia to decrease chances of prolapse, especially if considering laparoscopic surgery. No single intraoperative anesthetic agent or technique has demonstrated superiority. Adequate postoperative pain management is vitally important to avoid the detrimental effects of hypertension and tachycardia.
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PALAVRAS-CHAVE
Síndrome de Marfan;
Anestesia geral;
Cuidados pré-operatórios

Síndrome de Marfan: novos critérios diagnósticos, mesma abordagem anestésica? Relato de caso e revisão

Resumo
Justificativa: A síndrome de Marfan (SMF) é uma doença do tecido conjuntivo que envolve principalmente os sistemas: cardiovascular, músculo-esquelético e visual. Os problemas mais graves incluem dilatação da raiz da aorta e dissecção. O manejo anestésico é vital para a melhora da morbidade perioperatória.
Relato de caso: Homem de 61 anos com SMF, apresentando-se principalmente com *pectus carinatum*, escoliose, ectopia da lente, pneumotórax e espontâneo anterior e aneurisma da aorta e dissecção, submetido à colocação de prótese aórtica toracoabdominal. O paciente foi submetido à colecistectomia videolaparoscópica de rotina devido à litíase. Os achados importantes ao exame pré-operatório foram cifoescoliose tóracolombar e murmúrio metálico em exame cardíaco. A radiografia de tórax revelou ângulo de Cobb de 70° e o ecocardiograma mostrou evidência de prótese mecânica aórtica sem alterações.
Discussão: A avaliação pré-operatória deve ter como foco as anormalidades cardiopulmonares. O anestesiologista deve estar preparado para uma intubação potencialmente difícil. O posicionamento adequado e apoio para o membro antes da indução são fundamentais para evitar lesões nas articulações. Profilaxia antibiótica deve ser considerada para endocardite bacteriana subaguda. O paciente deve ser cuidadosamente posicionado para evitar lesões das articulações. O monitoramento cardiovascular é obrigatório no período intraoperatório: evitar manobras que podem levar à taquicardia ou hipertensão; controlar a pressão das vias aéreas para evitar pneumotórax e manter uma volemia adequada para diminuir as chances de prolapso, especialmente em caso de laparoscopia. Nenhum agente anestésico ou técnica demonstrou superioridade no período intraoperatório. O tratamento adequado da dor no pós-operatório é de vital importância para evitar os efeitos deletérios da hipertensão e taquicardia.
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Introduction

The MFS (MFS) is an autosomal dominant condition caused by a mutation in the FBN1 gene on chromosome 15 that encodes the protein fibrillin. This defect results in a set of expressions of various organs and systems, being musculoskeletal, cardiovascular and ophthalmic manifestations the most notorious.

It has an estimated incidence of 2–3 per 10,000 inhabitants.[1]

In 2010 the Ghent Nosology was revised, and new diagnostic criteria superseded the previous agreement made in 1996. The seven new criteria can lead to a diagnosis, being necessary to fulfill just one of the criteria:[2]

In the absence of a family history:

1. Aortic root Z-score ≥2 + *ectopia lentis*
2. Aortic root Z-score ≥2 + FBN1 mutation
3. Aortic root Z-score ≥2 + systemic score >7 points
4. *Ectopia lentis* AND an FBN1 mutation with known aortic pathology

In the presence of a family history:

1. *Ectopia lentis*
2. Systemic score ≥7
3. Aortic root Z-score ≥2

Points for systemic score:

Wrist AND thumb sign = 3 (wrist OR thumb sign = 1)

Pectus carinatum deformity = 2 (*pectus excavatum* or chest asymmetry = 1)

Hindfoot deformity = 2 (*pes planus* = 1)

Dural ectasia = 2

Protrusio acetabuli = 2

Reduced upper segment/lower segment ratio AND increased arm/height AND no severe scoliosis = 1

Scoliosis or thoracolumbar kyphosis = 1

Reduced elbow extension = 1

Facial features (3/5) = 1 (dolichocephaly, enophthalmos, downslanting palpebral fissures, malar hypoplasia, retrognathia) Skin striae = 1

Myopia >3 diopters = 1

Mitral valve prolapse = 0.25

In 1972, the decrease in average life expectancy in these patients was due to the changes on cardiovascular system, aortic aneurysm rupture being the major cause of mortality.[3] In 2010 life expectancy for patients with MFS has increased >25% since 1972. Reasons to sustain this dramatic increase may include benefits arising from cardiovascular surgery, and greater proportion of milder cases due to increased frequency of diagnosis. Medical therapy (including beta blockers) was also associated with an increase in probable survival.[4]

The potential risk of cardiac and respiratory events in a patient with MFS justifies the importance of an opportune

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