Medical Treatment or Endovascular Stent-Graft Treatment for Acute Aortic Syndrome

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22.2 Pathogenesis of Aortic Dissection

Chronic hypertension affects the arterial wall composition, causing intimal thickening, fibrosis and calcification, and extracellular fatty acid deposition. Moreover, adventitial fibrosis may obstruct nutrient vessels feeding the arterial wall as well as small intramural vasa vasa, which may result in necrosis of smooth muscle cells and fibrosis of elastic structures rendering the vessel wall vulnerable to pulsatile forces and creating a substrate for aneurysms and dissections [1–11]. In addition to chronic hypertension, smoking and dyslipidemia and potentially the use of crack cocaine are modulating risk factors. On rare occasions, inflammatory diseases destroy the media layers and cause weakening, expansion and dissection of the aortic wall. Iatrogenic aortic dissection may occur in association with invasive retrograde catheter interventions, or during or after valve or aortic surgery [12–14]. Given the morbidity and mortality of iatrogenic aortic dissection careful assessment is strongly encouraged in patients with unexplained hemodynamic instability or malperfusion syn-
dromes following invasive vascular procedures or aortic surgery (Table 22.1).

Finally, pregnancy-related dissection although a dramatic scenario is a rare event as long as the patient is not affected by any form of connective tissue disease. The putative association of pregnancy in otherwise healthy women and acute dissection may largely be an artifact of selective reporting. Pregnancy is a common condition and may coincidentally occur only with concomitant existence of other risk factors such as longstanding or pregnancy-associated hypertension, or Marfan’s syndrome. Preliminary data from the International Registry of Aortic Dissection (IRAD) show that pregnancy in Marfan’s syndrome is not associated with aortic tears, unless root size exceeds 40 mm.

### 22.2.1 Marfan’s Syndrome

Among hereditary diseases, Marfan’s syndrome is the most prevalent connective tissue disorder with an estimated incidence of 1/7,000 and an autosomal dominant inheritance with variable penetrance. More than 150 mutations on the fibrillin-1 (FBN-1) gene have been identified encoding for a defective fibrillin in the extracellular matrix, which may affect the ocular, cardiovascular, skeletal and pulmonary systems, as well as skin and dura mater. The diagnosis of Marfan’s syndrome is currently based on revised clinical criteria of the Gent nosology [15]. The Gent criteria pay particular attention to genetic information like Marfan’s syndrome in kindred of an unequivocally affected individual. Moreover, both skeletal and cardiovascular features are major (e.g., diagnostic) criteria if four or more of eight typical manifestations are present. Considering, however, borderline manifestations such as the MASS phenotype (mitral valve, aorta, skeleton, and skin), or subtle phenotypic features (“forme fruste”), the molecular analysis of suspected Marfan’s syndrome and the delineation of criteria for differentiating other inherited conditions (genotypes) from a Marfan phenotype are attracting interest [16–20]. The clinical variety of Marfan’s syndrome is only partially explained by the number of mutations on the FBN-1 gene. Genetic heterogeneity and the involvement of a second gene (Marfan syndrome type 2, MFS2) may further add to the broad spectrum of symptoms [21].

A common denominator of all phenotypic forms of aortic wall disease is the dedifferentiation of vascular smooth muscle cells not only with classic aneurysm formation, but also from enhanced elastolysis of aortic wall components [22], as shown in a fibrillin-q-deficient animal model [23]. Moreover, enhanced expression of metalloproteases in vascular smooth muscle cells of the aorta of Marfan patients may promote both fragmentation of medial elastic layers and elastolysis, thus initiating an activated phenotype of smooth muscle cells [24]. In parallel, expression of peroxisome proliferator-activated receptor-γ (PPAR-γ) is upregulated in smooth muscle cells of the aorta of Marfan patients and with cystic medial degeneration, and correlates with clinical severity, while vascular smooth muscle cell apoptosis is likely to be related to progression of aortic dilatation. Thus, PPAR-γ expression might reflect the pathogenesis of cystic medial degeneration and disease progression in the aorta of Marfan and non-Marfan patients without any vascular inflammatory response [25].

### 22.2.2 Ehlers-Danlos Syndrome

Ehlers-Danlos syndrome (EDS) is a heterogeneous group of heritable connective tissue disorders characterized by articular hypermobility, skin hyperextensibility and tissue fragility. Eleven types of EDS have been characterized; the true prevalence of EDS is unknown. An aggregate incidence of 1/5,000 births is often cited with no racial or ethnic predisposition. Aortic involvement is seen primarily in autosomal dominant EDS type IV [26].

### 22.2.3 Annuloaortic Ectasia and Familial Aortic Dissection

More than five mutations in the FBN-1 gene have now been identified in patients presenting with either sporadic or familial forms of thoracic aortic aneurysms and dissection [27, 28]. Histological examination of the aortic wall reveals elastolysis or loss of elastic fibers, deposits of mucopolysaccharide-like materials and cystic medial degeneration similar to Marfan’s syndrome. However, no abnormalities of types I and III collagen or any specific fibrilopathy were found in fibroblast cultures.