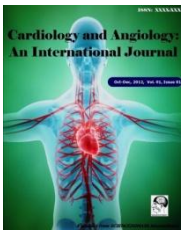

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Aortic Pathologies and Pregnancy: A Special Focus on Connective Tissue Disorders

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Authors' contributions

This work was carried out in collaboration between all authors. Author PB drafted the manuscript and mentored the writing process. Authors LG, MG and XB reviewed the literature and corrected the initial draft. Author GV wrote the manuscript, performed the corrections suggested from reviewers and selected the quoted references. All authors read and approved the final manuscript.

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ABSTRACT

Pregnancy represents a physiological status which, due to clear and typical hormonal and hemodynamic changes, is frequently accompanied by a high morbidity of aortic structures, in all of its segments. Such morbidity might become patent when a pre-gestational situation exists, especially with the mother suffering from connective tissue disorders. Nevertheless, morbid occurrences that will be met only during pregnancy are well known, with particular pathophysiological and etiological theories, as well as a diversity of treatments proposed in such an unusual setting. The authors discuss the main pathologies of aorta that are seen during pregnancy, from a theoretical point of view, and from a historical perspective as well. A special focus is made to the connective tissue disorders, and the theoretical considerations are illustrated with images of dissecting aneurysms of aorta.

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1. INTRODUCTION

Medical literature offers a diversity of situations related with pregnancy and specific aortic pathologies. During pregnancy, the heart undergoes different functional changes, while the entire cardiovascular system adapts to the new physiological situation, with increases in cardiac output, arterial compliance, and extracellular fluid, etc. [1]. Maternal blood volume may increase by up to about 30 per cent of the levels prior to pregnancy [2].

If physiological changes are almost clearly defined and extensively cross-examined, heart disease and pregnancy as a correspondence to each-other will still raise controversies. It is believed that cardiac disease will affect approximately two per cent of pregnancies in the Western world, with authors expanding this figure from a minimum to 0.2% to a maximum of 4% [3,4]. Unfortunately, both these data are from developed and industrialized countries: little is known from third-world and low-income countries. Albeit lack of global data, aorta and its valvular system has been of particular interest during pregnancy, since maternal mortality might achieve figures as high as 40%, especially when aortic stenosis coexists with mitral stenosis [5].

2. THEORETICAL AND HISTORICAL CONSIDERATIONS

There is a wide casuistics related with pregnancy and specific aortic pathologies. Mostly referred are the rupture and aortic dissection; aortic dilatation; Marfan syndrome and its foreseeable complications during gestation, as well as other conditions. The rupture of the sinus of Valsalva during pregnancy has been as well reported during pregnancy [6].

Nevertheless, mostly quoted are complications related with aortic coarctation, rupture and dissection of aorta during pregnancy, of traumatic or non-traumatic origin. Yentis et al when discussing the pregnancy and coarctation of aorta suggest that vaginal delivery with regional analgesia might have a good outcome, differing from other sources that are more prone to ‘elective’ cesarean section [7]. A highly morbid situation, but rather infrequent, is the recurrent aortic coarctation. Sources refer such a recurrence during pregnancy in previously surgically repaired aortic coarctation [8].

Dissecting aneurysms of aorta have been classified from Stanford according to a simple anatomical criterion, namely the involvement or not of the ascending aorta [9]. Thus, a dissecting aneurysm that involves the aortic root and the ascending aorta is classified as type A (Fig. 1); type B dissections do not involve the ascending aorta (Fig. 2).

Aortic rupture in the background of an anamnestic silent post-traumatic aneurysm have been reported with the patient surviving fifteen years with an aneurysm after a road traffic accident [10]. Here again, pregnancy was imputed for the rupture of a silent post-traumatic aneurysm. Dissection of aorta is another issue of major concern, especially when it happens in a total absence of risk factors. Such a spontaneous dissection in a previously healthy pregnant woman, with fatal outcome, suggests again the importance of raising awareness for aortic events in pregnancy [11]. The highly lethal event of spontaneous aortic dissection as a rule will be related to a positive history for connective tissue disorders, or other pathologies [11,12].

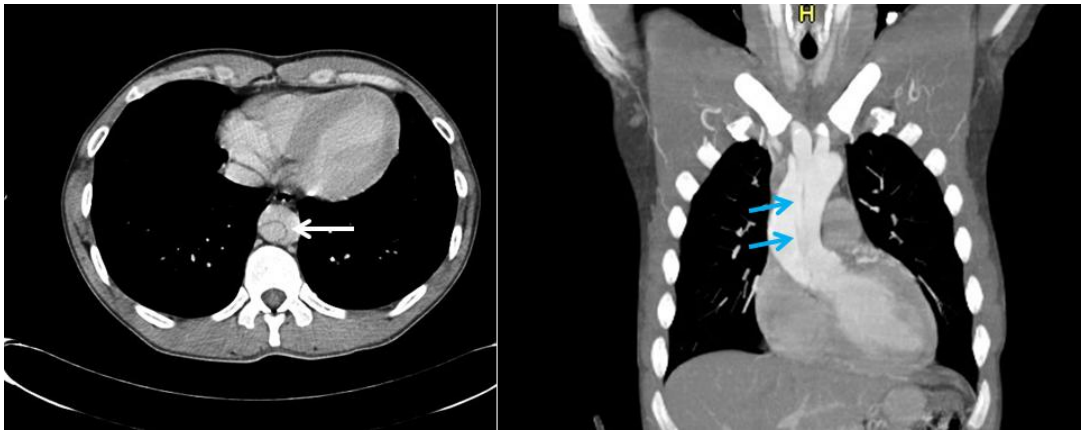

Fig. 1. Type A Stanford dissecting aneurysm of aorta. Note the double lumen in the axial image of angiographic CT (white arrow; left part of the image) and the double contrasting in the sagittal reconstruction view (blue arrows; right part of the image)

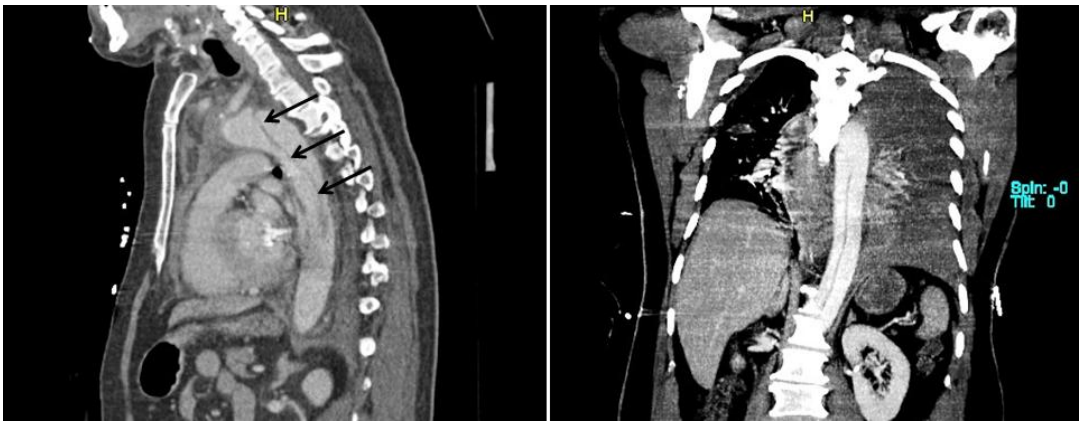

Fig. 2. Type B Stanford dissecting aneurysm of aorta. Note the dissection of the thoracic aorta (black arrows, sagittal reconstruction view; left part of the image). The dissection starts immediately distal to the origin of the left subclavian artery, without involving the ascending aorta

Aortic pathology in pregnancy is a relatively recent theme in the specialized literature. In fact, the first case of aortic dissection in a primiparous woman has been reported only in 1949, with a fatal case with an extension of the dissecting vessel reaching the renal artery [13]. Almost a decade later, dissecting aortic aneurysms were reported in multiparous women from two sources [14,15]. All these remote and initial case reports, were constantly related with the presence of Marfan's syndrome. Thus, particular importance is given to other body stigmata specifically related with this rare syndrome, such as arachnodactyly, kyphoscoliotic alterations of the skeleton and maternal tall stature; although these features are not constantly reported.

Several reviews and detailed descriptions of Marfan's syndrome, with diagnostic criteria, separated in major and minor criteria, as well as divided accordingly with the interested

organs or body systems, are available [16-19]. The Table 1 summarizes some of these major and minor diagnostic criteria, with the typical findings of Marfan's syndrome, in the cardiovascular system, ocular and skeletal system, and other particularities.

Table 1. Diagnosis of Marfan's syndrome (Adapted from 13,14,16-19)

Features, organs and systems involved in the Marfan's syndrome	Major/minor diagnostic criterion (quoting sources)
Heart and vascular system	
Aneurysm of the ascending aorta involving at least the sinuses of Valsalva	Major criterion [17]
Dissection of the ascending aorta	Major criterion [17]
Mitral valve prolapse	Minor criterion [17]
Dilatation of the main pulmonary artery; younger than age 40 years	Minor criterion [17,19]
Calcification of the mitral annulus; younger than age 40 years	Minor criterion [17,19]
Dilatation or dissection of the descending thoracic or abdominal aorta; younger than age 50 years	Minor criterion [17;19]
Dilatation of the ascending aorta	[18]
Haemopericardium	[16]
Skeletal changes	
Pectus carinatum/pectus excavatum	Major criterion [17,16]
Upper to lower segment ratio <0.85 or arm-span to height ratio >1.05	Major criterion [17]
Pes planus	Major criterion [16,17]
Scoliosis >20° or spondylolisthesis; kyphoscoliosis	Major criterion [17,16]
Extension at elbows<170°	Major criterion [17,19]
Protrusio acetabuli (ascertained on radiographs)	Major criterion [17,19]
Arachnodactyly	[16,18]
Winged scapula	[14]
Facial features	
Retrognathia; enophthalmos; dolichocephaly; malar hypoplasia.	Minor criterion [17,19]
Ocular system	
Ectopia lentis	Major criterion [17-19]
Flat cornea	Minor criterion [17,19]
Increased axial length of the globe	Minor criterion [17,19]
Hypoplastic iris or hypoplastic ciliary muscle	Minor criterion [17,19]
Other organs/systems	
Pulmonary system (Spontaneous pneumothorax; apical blebs)	Minor criterion [17,19]
Dura mater (Lumbosacral dural ectasia)	Major criterion [17,19]
Kidney (Renal cortical necrosis)	[13,16]
Skin (Striae distensae/atrophicae)	Minor criterion [17,19]

The human intuition and medical casuistics are far more remote than cases reported from the twentieth century, with regard to aortic involvement during pregnancy, and the severity of such a condition. In a very interesting historical review, Mazzotti et al. [20] describe their own case, of a young pregnant woman dying from a cardiac congenital disease. Their diagnosis was made post mortem in a wax model of the young woman, more than two centuries from

her passing away. This artistic masterpiece of a human wax model was produced in 1782, showing an open arterial duct; thus proving that awareness for congenital heart diseases, and their importance for the outcome of a pregnancy, is of a very old date. As an artistic and meticulous reproduction of cadaveric findings, this wax model of the Italian sculptor Sussini is a touchable proof of the risks that pregnancy might bring for the cardiovascular system in general and for the aorta in particular.

3. CONCLUSION

Pregnancy is a physiological condition that predisposes the installation of unfavorable hemodynamic changes vis-à-vis the heart and the aorta, especially in the background of previous positive history for cardiac pathologies, even if these have a genetic character or not. Hypertensive disorder in pregnancy and pre-eclampsia might be the two most frequent pathophysiological mechanisms.

Authors suggest that pregnancy might contribute in the rupture of arterial aneurysms [21]. In fact, starting from the third month of the pregnancy till the seventh of the latter, circulation will undergo a physiological acceleration, with an increase in the blood volume and in the cardiac stroke as high as 30-50%. Hypertensive complications during pregnancy will be accompanied with systemic endothelial dysfunction, impairing substantially the physiological vasodilatation [22]. In the other hand, blood pressure will present unexpected changes starting from the seventh month of pregnancy, and such a period will result critical for patients suffering from coarctation of aorta, or from other cardiac and circulatory conditions.

A careful selection of cases that should avoid vaginal delivery and perform parturition through cesarean section is always warranted, mainly with the aim of avoiding Valsalva maneuvers, which increases substantially intrathoracic pressures [23,24]. Of course, it is advisable a surgical correction of all previously diagnosed conditions (coarctation of aorta etc.) before progeny conception. A vaginal delivery, probably combined with epidural analgesia, is anyway very much possible for pregnant women with cardiac risk factors, here including aortic pathologies, in facilities that offer emergency cardiac surgery; and positive outcomes have been constantly referred [7,25].

Aortic pathologies have high mortality rates, and prevention is obviously very important; thus a high clinical index of suspicion is important [26]. Detailed guidelines for the diagnosis and treatment of the aortic dissection are available [27]. Authors emphasize the role of other inherited disorders such as Ehler-Danlos syndrome and familial forms of thoracic aortic dissection and aneurysm; the concept of ageing of aorta is included and normal aortic dimensions are provided in tables, as well as the indications for surgery [27]. Other vasculitic disorders that affect major vessels might involve aorta as well [28]. The bulk of the casuistics and of the experimental work is naturally concentrated on the prevention strategies, early diagnosis and interventional options. This acquired knowledge will help clinicians toward a better management of a highly serious occurrence, such as of the aortic pathologies during pregnancy, especially in women with a positive history for connective tissue disorders.

CONSENT

Not applicable.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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