Clinical Guideline

Management of Aortopathy in Pregnancy

SETTING	**University Hospitals Bristol**
GUIDELINE FOR	The cardiac obstetric multidisciplinary team
PATIENT GROUP	Pregnant women with dilated aortas, including those with Marfan's Syndrome (MFS), Turner's Syndrome (TS) and related connective tissue diseases (CTDs) causing aortopathy, bicuspid aortic valve (BAV) with associated aortopathy, post Ross operation, or congenital heart repair associated with enlargement of the ascending aorta.

GUIDANCE

Pregnant women with dilated aortas, particularly those with Marfan's Syndrome (MFS), Turner's Syndrome (TS) and other inherited aortopathies, such as those due to mutation of MYH11, ACTA2 or TGFBR2, are at risk of aortic dissection in pregnancy and in the post- partum period due to hormonal effects on elastin. The highest risk period is in the third trimester, peripartum and for several days post-partum. For the purposes of this document, MFS refers to Marfan's Syndrome and those conditions with a high risk of aortic dissection as above.

The risk has been studied most in patient with MFS and BAV:-

In MFS, dissection risk is increased in women with:

- a family history of dissection;
- previous aortic dissection or aortic surgery;
- aortic regurgitation; or
- a rapidly growing aorta.

Risk is reduced by beta-blockade.

Dissection is unlikely in pregnancy if the aorta measures <45mm* and none of the above features are present. However, the clinician must be mindful that dissection can still occur and Type B dissection in particular, cannot be predicted by aortic root size.

All patients with Marfan's syndrome should be considered high-risk pregnancies and be managed in a cardiac surgical centre. Vaginal delivery with a passive second stage of labour is the aim, unless the aortic root measures >45mm, in which case elective Caesarean section for cardiac reasons is recommended. Trans-thoracic echocardiograpy should be undertaken at least 6 weekly until 6 weeks post-partum. All patients should have blood pressure carefully controlled, ideally with beta-blockers if tolerated and undergo 4 weekly growth scans from 26 weeks gestation (due to beta-blocker associated IUGR).

BAV aortopathy is relatively common and the risk of dissection is lower with few dissections reported in registries and one prospective longitudinal study. The ascending aorta is typically dilated, rather than the aortic root. Pregnancy should be avoided if the aorta measures >50mm. As with MFS blood pressure control and surveillance of the aorta by echocardiography is recommended, though 6-8 weekly echoes are sufficient unless the aorta is >45mm. Vaginal

delivery should be the aim and elective Caesarean section for cardiac reasons is not necessary.

Patients can be risk-stratified as follows,

EXTREMELY HIGH RISK: MFS with Sinus of Valsalva measuring >45mm
MFS with a growing sinus of Valsalva
MFS with previous surgery/dissection

VERY HIGH RISK: MFS with Sinus of Valsalva measuring 40-45mm
MFS with aortic regurgitation
MFS with family history of dissection

HIGH RISK: MFS with Sinus of Valsalva measuring <40mm
BAV aortopathy with ascending aorta >45mm

MODERATE RISK: BAV aortopathy with ascending aorta 40-45mm

LOW RISK: BAV aortopathy with ascending aorta <40mm**The following table is a guideline for the management of these patients.**

	Beta-block	**Frequency of scan**	**Elective epidural**	**Second Stage**
MFS	All	4 weekly	All	Elective Caesarean section if **EXTREMELY HIGH RISK,** consider in **VERY HIGH RISK** Passive for VERY HIGH RISK and HIGH RISK
BAV with aorta>50mm	Yes	4 weekly	All	Passive
BAV with aorta 45-50mm	Yes	4-6 weekly	All	Semi passive with passive descent and up to 30 minutes pushing
BAV with aorta 40-45mm	No	6-8 weekly	Low threshold	Management of second stage as per usual obstetric guidelines
BAV with aorta <40mm	No	8-12 weekly	No	Management of second stage as per usual obstetric guidelines

Patients who have had the **Ross operation** or other repaired congenital heart defects associated with an enlarged aorta should be treated as for BAV aortopathy. Other thoracic aortic aneurysm syndromes should be treated as for MFS by a specialist team.

BAV +/- BAV aortopathy is common in TS. Pregnancy can occur in mosaic TS and with assisted conception. Death from aortic dissection has been reported in up to 2% pregnancies, though there is little evidence to guide management. Dissection can occur in the absence of hypertension or congenital heart disease. All patients should be treated as BAV with with aorta >50mm with delivery in a surgical centre. Those with CHD or hypertension should be treated as MFS as above with beta-blockade and consideration of elective Caesarean section.

*Aortic measurements should be indexed to body surface area. This is especially important in TS.

RELATED DOCUMENTS Regional Referral Pathway

REFERENCES

Anderson RA, Fineron PW, BJOG 1994 Dec; 101 (12); 1085–1088
Donnelly, RT, et al. JACC 2012, 60(3):224
Goland S, Elkayam U, Circ 2009 Feb 3;119(4):619-23
Immer FF Ann Thorac Surg 2003 Jul;76(1):309-14
Lind J, Wallenburg HC, Eur J Obs Gynaecol Reprod Biol 2001;98(1):28-35
Lipscomb KJ et al BJOG 1997;104:201-6
McKellar SH et al. Am J Cardiol. 2011 Jan;107(1):96-9.
Meijboom LJ et al, EHJ 2005;26(9):914-20
Mulder BJ, Meijboom LJ. J Am Coll Cardiol.2012 17;60(3):230-1.
Pyeritz RE, Am J Med 1981 Nov;71(5):784-90
Rossiter JP et al, AmJOG 1995;173;1599-1606
Carlson M, Airhart N, Lopez L, Silberbach M. Circulation. 2012 Oct 30;126(18):2220-6.

SAFETY **Careful management with an experienced multi-disciplinary team is advised with an individual care plan for each woman**

QUERIES Contact any of the following via switchboard

Dr S Curtis, Consultant Cardiologist
Dr G Stuart, Consultant Cardiologist
Dr Y Ismail, Locum Consultant Cardiologist
Miss J Trinder, Consultant Obstetrician
Miss A Mohan, Consultant Obstetrician
Miss L Ashelby, Consultant Obstetrician