# Fatal & Rare Combination Of Stanford Type A-Aortic Dissection, Bicuspid Aortic Valve & Pericardial Tamponade-A Case Report.

**Dr Rosemol Xaviour[*], Dr Girijamony V.K[**], Dr Joy Augustine[***], Dr.Simi .S[****]**

[*] Junior Resident, Government Medical College, Thrissur, Kerala, India.
[**] Professor, Department Of Anatomy, Government Medical College, Thrissur, Kerala, India.
[***] Professor, Department Of Pathology, Government Medical College, Thrissur, Kerala, India.
[****] Senior Resident, Department Of Pathology, Government Medical College, Thrissur, Kerala, India.

***Abstract-*** A 40 year old female was brought for post mortem examination with history of epigastric pain of one day duration followed by death after hospitalisation on the very same day. She was unmarried & had mild mental retardation but she was capable of doing household chores. She also had recurrent episodes of epigastric pain for last 1 year for which she was treated with antacids. On examination pericardial tamponade was found as the cause for death. Detailed examination of heart & histology revealed associated bicuspid Aortic valve & Stanford type A aortic dissection.

***Index Terms***- bicuspid Aortic valve epigastric pain, mental retardation, pericardial tamponade, Stanford type A aortic dissection

## I. INTRODUCTION

When sudden death (SD) occurs in adults and elderly persons, coronary atherosclerosis is the usual cause. On the contrary, a large spectrum of cardiovascular diseases, both congenital and acquired, may account for SD in the young. These diseases are frequently concealed and discovered with surprise only at autopsy by means of a thorough macroscopic and microscopic investigation.[1]

## II. CASE HISTORY

A 40 year old female was hospitalised with vague epigastric pain of one day duration and died on the very same day. She was unmarried, had mild mental retardation but was capable of doing routine household chores. On post-mortem examination she was normally built and nourished. External features were not pointing towards any chromosomal anomaly. Internal examination revealed pericardial tamponade with about 1.5 litres of blood collected in the pericardial cavity which was fatal enough to cause death of the patient.

On detailed examination, the aortic valve was bicuspid and the aorta showed dissection staring from the root of Aorta up to the termination of aortic arch[figure 1].The coronaries were patent and there were no associated anomalies. On microscopy, the aorta showed dissection extending between media and adventitia [figure 2 & figure 3]

## III. DISCUSSION

Most patients with a bicuspid aortic valve are unaware of the diagnosis until late in life because symptoms and physical findings often are absent for many years[2] . The tissue abnormality in patients with a bicuspid aortic valve is not confined to the valve leaflets; these patients are at increased risk of aortic aneurysm and dissection. Intrapericardial rupture of aortic dissecting aneurysm may occur in some which may prove fatal [3,]Thoracic aorta dissection occurs as blood flow is redirected from the aorta (true lumen) through an intimal tear into the media of the aortic wall (false lumen)[4,5]. A dissection plane that separates the intima from the overlying adventitia along a variable length of the aorta is created within the media. At the tissue level, the aorta shows cystic medial necrosis, loss of elastic fibres, increased apoptosis, and altered smooth muscle cell alignment[2,11] All dissections that involve the ascending aorta are grouped together as Stanford type A, regardless of where the primary tear occurs and Stanford type B involves only the descending Aorta[4,8,9,10]The risk of aortic dissection in patients with a bicuspid valve is 5 to 9 times higher than in the general population[2] The association of bicuspid aortic valve with aortic aneurysm and dissection suggests the possibility that a bicuspid valve, at least in some patients, is only the most identifiable manifestation of a systemic connective tissue disorder[6,7]

William's syndrome is an autosomal dominant disorder that is characterized by supravalvular aortic stenosis, peripheral pulmonary stenosis, obstructive coronary lesions, abnormal facies and mental retardation. This syndrome was found associated with a deletion of a region of the same chromosome 7 (7q11.23)[1] In addition, a bicuspid aortic valve is present in 10% to 12% of women with Turner syndrome.[2] Typical features of Turner syndrome (eg, short stature in females with webbed neck and broad chest) or Williams syndrome (e.g., elfin facies, mild retardation) may suggest the possibility of bicuspid aortic valve[3,12] . Type IV Ehlers-Danlos syndrome is a connective tissue disorder of the pro$\alpha$1(III) chain of Type III collagen with an incidence of 1 in 5000. The structurally abnormal media is susceptible to dissection. There are also familial aggregations of dissection without discernable biochemical or genetic abnormalities[4]

## IV. CONCLUSION

A careful history and physical examination are essential in cases where the cause of abdominal pain seems obscure[1,13] Clinicians should think of common conditions first, but when developing a differential diagnosis they must consider risk factors and co morbid illnesses that predispose patients to uncommon entities. A case of mental retardation should always be thoroughly investigated since valvular anomalies and cardiac malformations usually coexist as part of syndromes and which may prove fatal as we have seen in the above case.

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## AUTHORS

**First Author** – Dr.ROSEMOL XAVIOUR, MBBS, Junior Resident, Department Of Anatomy, Government Medical College Thrissur, Kerala, India. Member Of Society of Clinical Anatomists and Indian Medical Association, email address-rosejos06@gmail.com,rosejos@rediffmail.com
**Second Author** – Dr. DR GIRIJAMONY V.K ,MBBS, MS, Professor & Head Of The Department Of Anatomy, Government Medical College Thrissur, Kerala, India., email address-girijamonyanatomy@gmail.com
**Third Author** – Dr. JOY AUGUSTINE ,MBBS, MD, Professor & Head Of The Department Of Pathology, Government Medical College, Thrissur, Kerala, India., Email – joyajoy@rediffmail.com
**Fourth Author** – DR.SIMI .S ,MBBS, MD, Department Of Pathology, Government Medical College, Thrissur, Kerala, India. Email-simi.0903@gmail.com

**Correspondence Author** – Dr.ROSEMOL XAVIOUR, MBBS ,Junior Resident, Department of Anatomy, Government Medical College Thrissur, Kerala, India., Email address-rosejos06@gmail.com, rosejos@rediffmail.com, Contact Number-9947269581

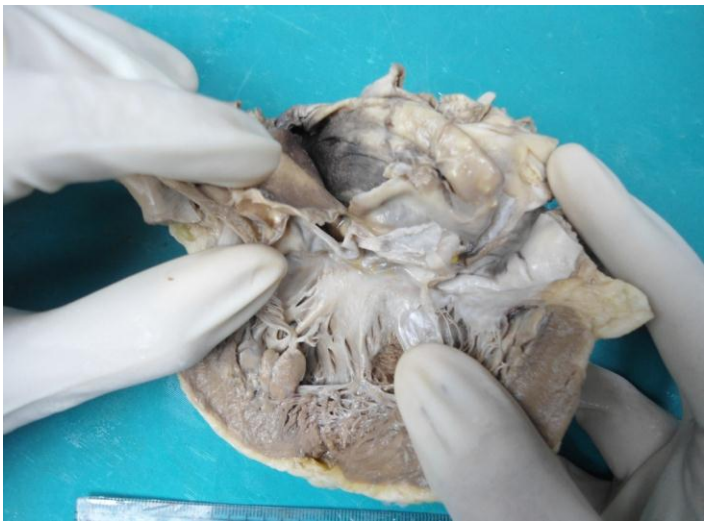

**Figure 1-The heart shows bicuspid Aortic valve & the plane of dissection starting from the root of Aorta**

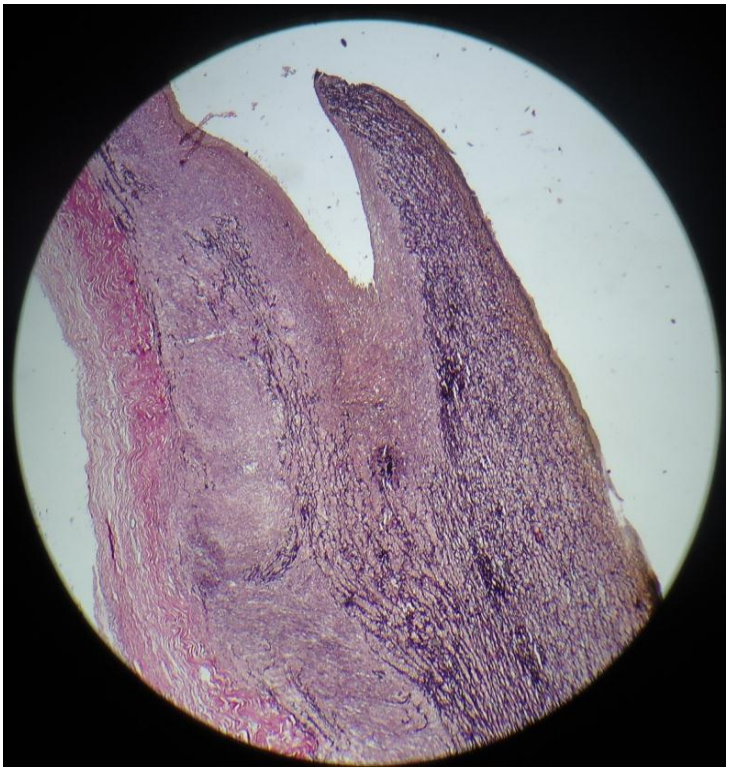

**Figure 2-Dissection extending from tunica intima to tunica media & tunica adventitia[H & E stain]**

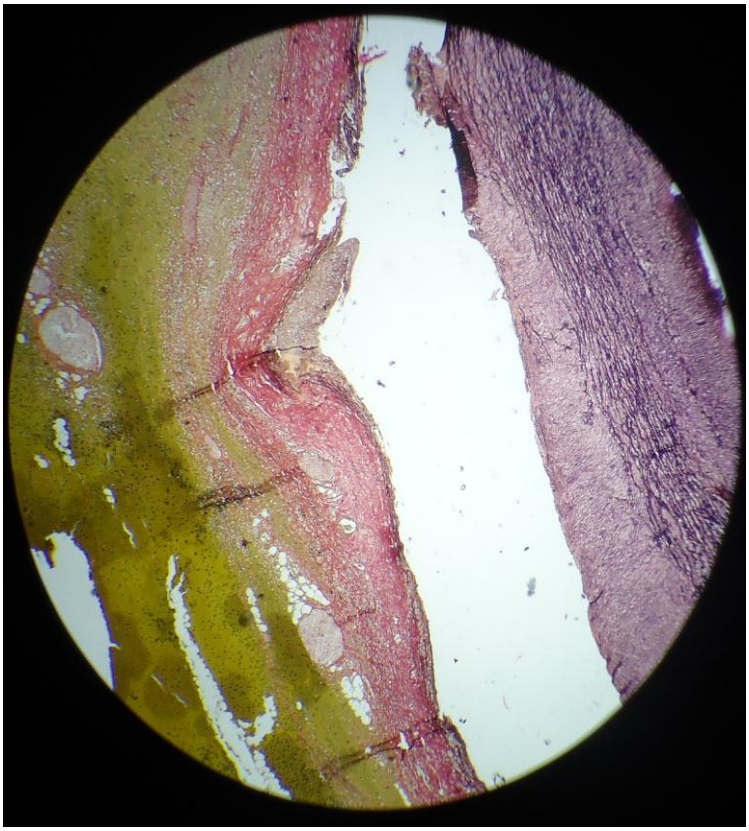

**Figure 3-Plane of dissection between Tunica media & adventitia[vangieson stain]**