Surgery for Cardiac Myxomas

INTRODUCTION

- Primary cardiac tumors- infrequent (0.0001%-0.5%) in autopsy series
- 40%-50% are myxomas
- Incidence of myxoma 0.5-1 case per million people per year
- Higher prevalence in women
- Observed in all age groups, mean age 42-69 years old

Schaff HV et al Sem Thorac Cardiovasc Surg 2000, Samanidis G et al Kardiologia Polska 2020

INTRODUCTION

- Most common location left atrium 72%-92%

(fossal ovalis in interatrial septum with or without a broad base)

- Right atrium 0.7%-7.5%
- Right ventricle 0.7%-2.5%
- Left ventricle 0.7%-3.6%
- Heart valve (aortic, mitral tricuspid) < 1%

Schaff HV et al Sem Thorac Cardiovasc Surg 2000, Samanidis G et al Kardiologia Polska 2020

CLINICAL PRESENTATION

- Asymptomatic patients - 3.2%-46.4%
- Systemic effects- 16.9%-32.4% (anemia, fever,weight loss, fatigue, arthralgia myalgia, Raynaud phenomenon, IL-6↑,immunological alterations)

- Obstructive symptoms (dyspnea, palpitations, tachycardia, AF, syncope, sudden death)
- Embolization- pulmonary, systemic, 30%-60% (CNS, upper/lower extremity, ACS mesenteric ischemia)

Schaff HV et al Sem Thorac Cardiovasc Surg 2000, Samanidis G et al Kardiologia Polska 2020

DIAGNOSIS

- Clinical examination/electrocardiography
- Differential diagnosis (benign or malignant tumors-primary or metastatic, thrombi, vegetations)
- TTE/TEE- 90% to 96% accurancy in diagnosis (nature, structure, mobility)
- Chest CT/Cardiac MRI/PET- coexisting conditions,staging
- Coronary angiography/CCT angiography
- Genetic analysis (familial and Carney complex form)

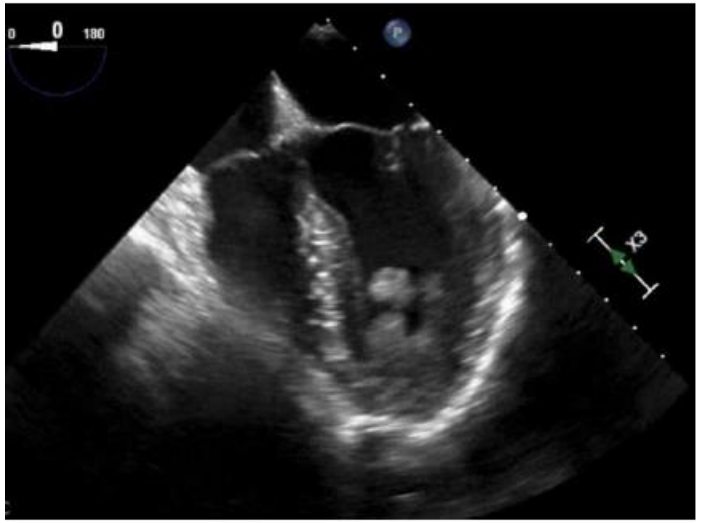
Schaff HV et al Sem Thorac Cardiovasc Surg 2000, Samanidis G et al Kardiologia Polska 2020

DIAGNOSIS

- Familial form- 5% of patients with cardiac myxoma
- Multiple myxomas/atypical location/younger patients/recurrence
- Mutation of the PRKAR1A gene

  (inactivation of PRKAR1A- ↑sensitivity and activation of protein kinase A by cAMP, promoting myxomatumorigenesis)

Schaff HV et al Sem Thorac Cardiovasc Surg 2000, Samanidis G et al Kardiologia Polska 2020, Gerhold C et al



CARNEY COMPLEX

- Described by Carney JA in 1985
- Exact prevalence unknown/750 cases since 1985/↑recurrence
- Decreased life span (57% of the deaths due to heart related causes)
- Inherited in autosomal dominant pattern
- Variable phenotype: cardiac myxoma, skin myxoma, myxoid mammary fibroadenomas, spotty skin pigmentations(lentigo,nevi), Cushing syndrome, pituitary adenoma, testicular tumors (Sertoli cell tumors)

Schaff HV et al Sem Thorac Cardiovasc Surg 2000, Samanidis G et al Kardiologia Polska 2020, Stratakis et al J Clinical Endocrinology Metabolism 2001

CARNEY COMPLEX

TABLE 4. Diagnostic criteria for CNC[a]

1. Spotty skin pigmentation with a typical distribution (lips, conjunctiva and inner or outer canthi, vaginal and penile mucosa)
2. Myxoma (cutaneous and mucosal)[b]
3. Cardiac myxoma[b]
4. Breast myxomatosis[b] or fat-suppressed magnetic resonance imaging findings suggestive of this diagnosis[c]
5. PPNAD[b] or paradoxical positive response of urinary glucocorticosteroids to dexamethasone administration during Liddle's test[d]
6. Acromegaly due to GH-producing adenoma[b]
7. LCCSCT[b] or characteristic calcification on testicular ultrasonography
8. Thyroid carcinoma[b] or multiple, hypoechoic nodules on thyroid ultrasonography, in a young patient
9. Psammomatous melanotic schwannoma[b]
10. Blue nevus, epithelioid blue nevus (multiple)[b]
11. Breast ductal adenoma (multiple)[b]
12. Osteochondromyxoma[b]

Supplemental criteria:

1. Affected first-degree relative
2. Inactivating mutation of the *PRKAR1A* gene

[a] **To make a diagnosis of CNC, a patient must either: 1) exhibit two of the manifestations of the disease listed, or 2) exhibit one of these manifestations and meet one of the supplemental criteria (an affected first-degree relative or an inactivating mutation of the PRKAR1A gene).**

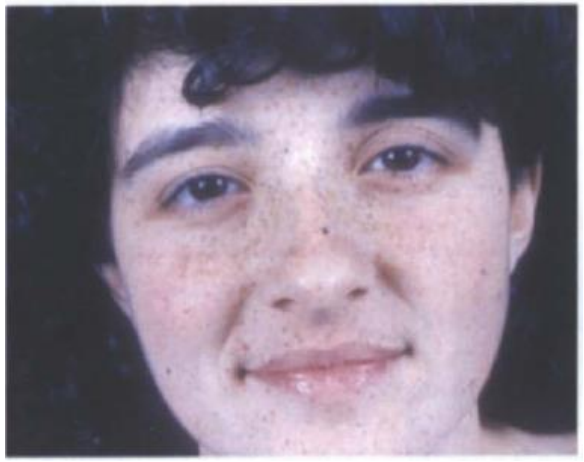
[b] With histologic confirmation.

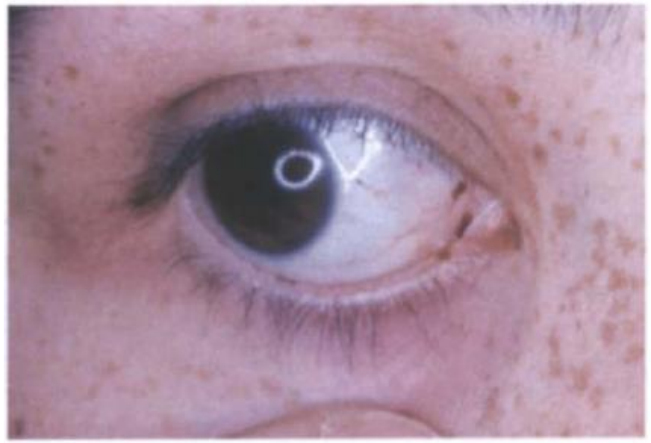
[c] See Ref. 40.

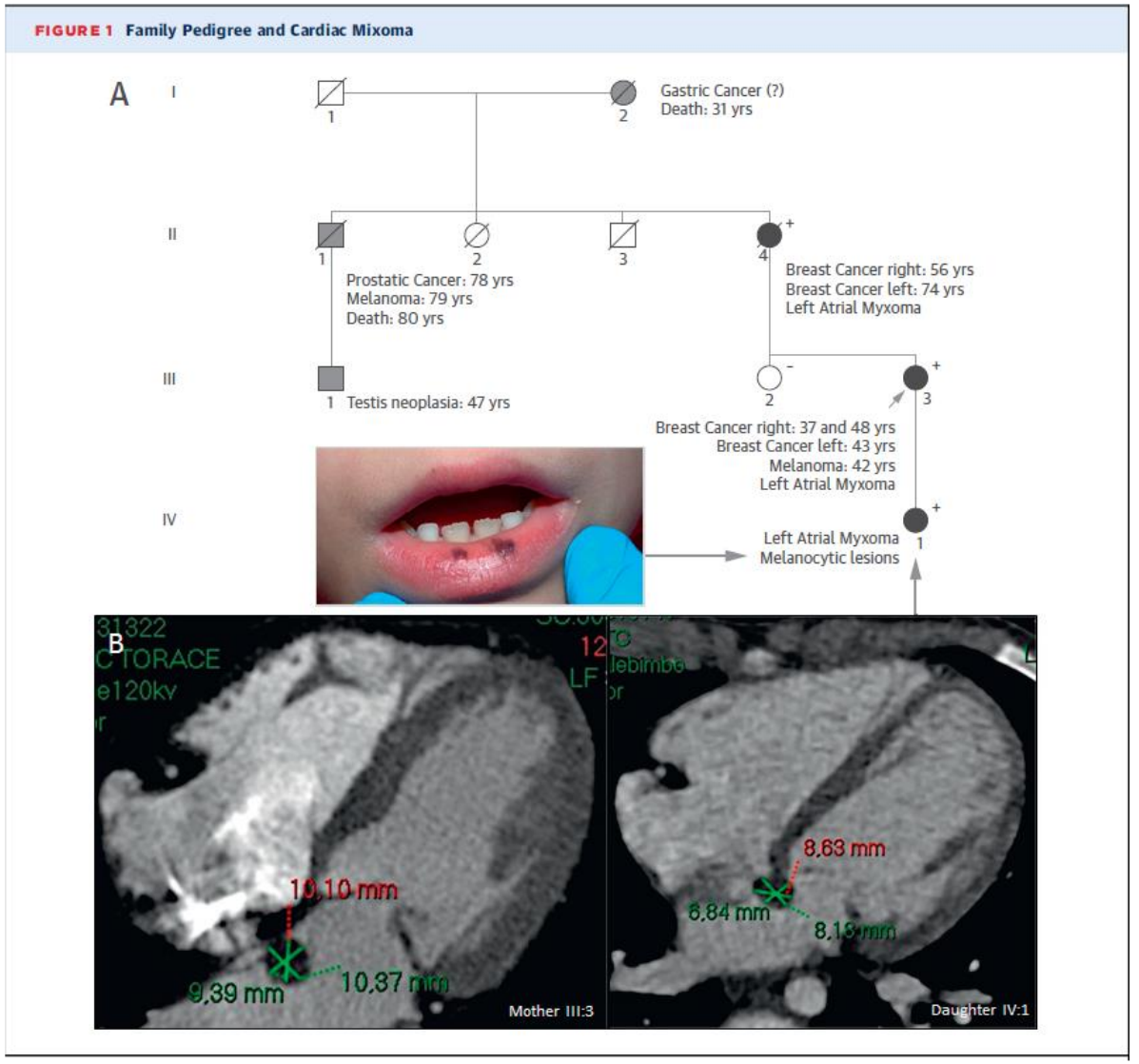
[d] See Ref. 10.

Stratakis et al J Clinical Endocrinology Metabolism 2001

CARNEY COMPLEX







Serio A et al JACC 2016, Schaff et al Sem Thorac Cardiovasc Surg 2000

SHORT-TERM RESULTS

- Resection is associated with low rate of postoperative complications
- Postoperative period is usually uneventful
- AF most common postoperative complication
- 30 day mortality 0%-10%

Samanidis G et al Kardiologia Polska 2020

LONG-TERM RESULTS

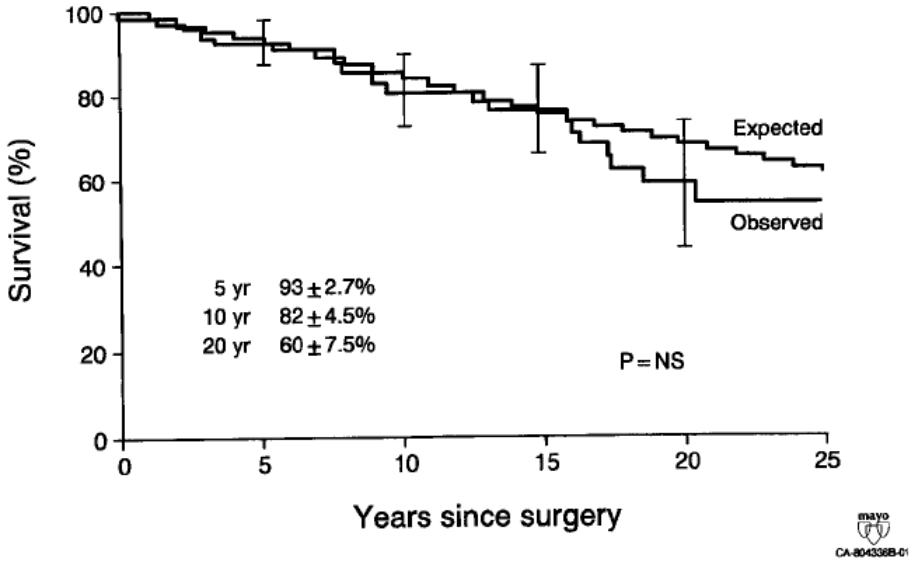


Figure 7. Comparison of survival rate of patients after myxoma removal with expected survival of the general population (1-sample log rank test).

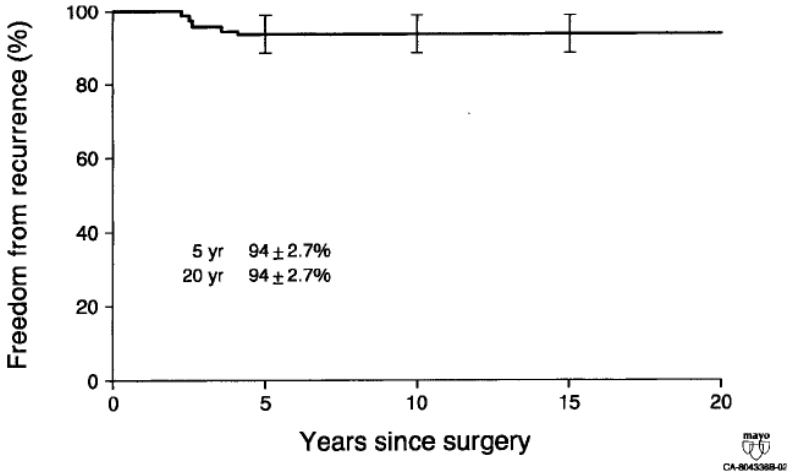


Figure 8. Survival free of recurrence of atrial myxoma.

Recurrence rate: 0%-7.4% (sporadic myxomas 1%-3%, familial disease 12%, Carney´s complex 22%)
Survival: 70%-96.8%

Schaff HV et al Sem Thorac Cardiovasc Surg 2000, Samanidis G et al Kardiologia Polska 2020

NÚSCH as RESULTS

- January 2010-December 2022 13402 on-pump procedures
- Surgery for cardiac myxoma 90 patients (0.67%)
- Urgent procedures, use of bicaval CPB/tumor resection
- 2 patients repair of iatrogenic ASD-pericardial patch repair
- Right atrial / 1 patient transaortic approach
- Median sternotomy 82/90 (81.1%)
- VATs 8/90 (8.9%)

MOST COMMON PREOPERATIVE SYMPTOMS

- **Systemic symptoms**: 8 (8,9%) (fatigue 6 (6,7%), fever 2 (2,2%))
- **Arterial emboli**: 11 (12,2%)
- **Dyspnea**: 19 (21,1%)
- **Chest pain**: 8 (8,9%)
- **Syncope**: 5 (5,6%)
- **Arrhythmia**: 7 (7,8%) (supraventricular tachycardia 3 (3,3%),
  PAF 4 (4.4%))
- **Asymptomatic**: 32 (35,6%)

PREOPERATIVE AND DEMOGRAPHIC CHARACTERISTICS

Variable n=90	N (%)
Age, years, mean±SD	59.4±13.5
Gender, male	30 (33.3%)
Euroscore II, mean±SD	2±2
NYHA class, mean±SD	1.5±1.2
Smoking	14 (15.6)
Diabetes mellitus	12 (13.3)
Hypercholesterolemia	33 (36.7)
Chronic renal disease	2 (2.2)
Atrial fibrillation	10 (11.1)
Coronary artery disease	9 (10)

PERIOPERATIVE AND POSTOPERATIVE CHARACTERISTICS

Variable n=90	n (%)
CPB, min, mean±SD	58.2±48.4
Aortic cross clamp, min, mean±SD	36.7±30
Bleeding,re-exploration	5 (5.6)
Stroke	1 (1.1)
Wound infection	2 (2.2)
Lung infection	1 (1.1)
Dialysis	0 (0)
Postoperative AF	22 (24,4)
Hospitalization, days, mean±SD	13±7.3
Hospital mortality	2 (2.2)
All-cause mortality, long-term	14 (15.6)

ASSOCIATED PROCEDURES

Precedure	n
Atrial septal defect repair	3
Tricuspid valve repair	10
MAZE procedure	9
CABG LAD (LIMA graft 2, SVG graft 3) RCA (SVG graft 2)	7
Mitral valve repair	6
Mitral valve replacement (mechanical valve)	1
VA-ECMO implantation	1
Septal myectomy	1
Replacement of the ascending aorta due to IMH	1
Aortic valve replacement (mechanical valve)	1

LOCATION OF CARDIAC MYXOMA

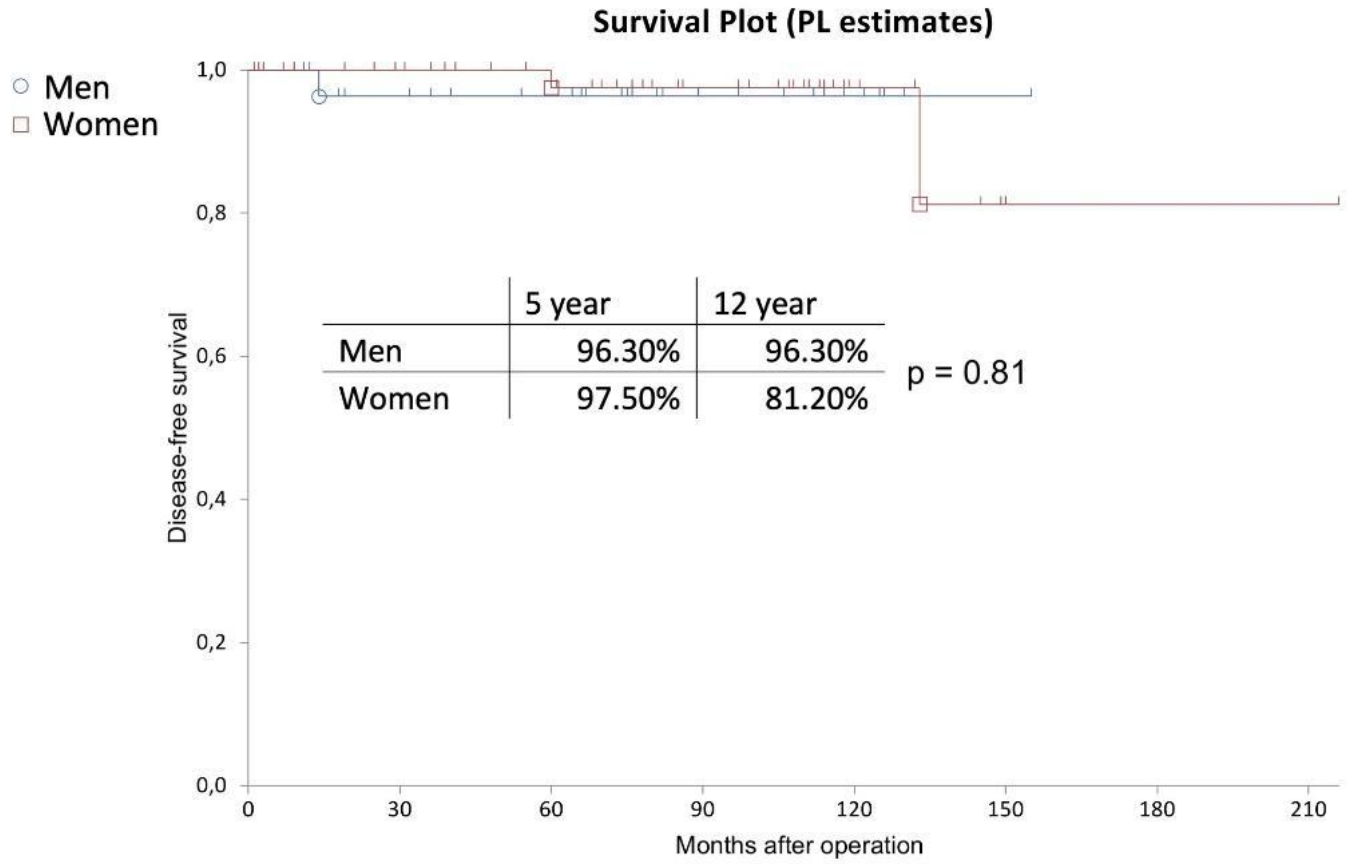
Location	n	%
Left atrium	**82**	**88.2**
Septum	68	73.1
Superior wall	3	3.2
Lateral wall	2	2.2
Posterior wall	1	1.1
Right/left superior pulmonary vein	2/1	2.2/1.1
Mitral valve annulus	5	5.4
Right atrium	**8**	**8.6**
Septum	5	5.4
Tricuspid valve (2 septal leaflet, 1PM)	3	3.2
Right and left atria	**1**	**1.1**
Septum	1	1.1
Left ventricle/ septum	**1**/1	**1.1**/1.1
Right ventricle/ apex	**1**/1	**1.1**/1.1

RECURRENCE

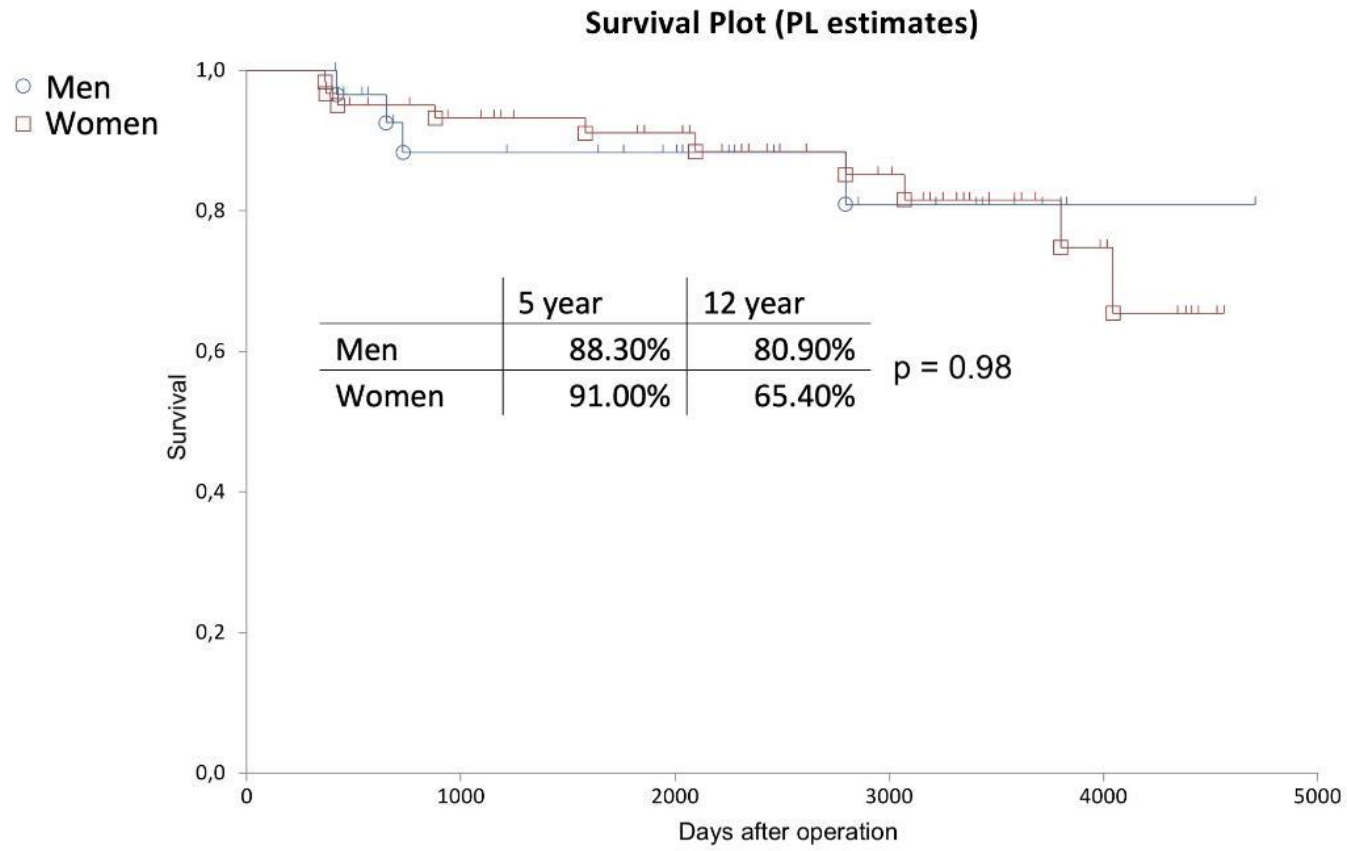
- **3 patients/90 (2,7%)**
- **mean time 55±19.7 months**

- 1 patient with Carney complex 2x after 48 months
- 1 patient sporadic myxoma after 88 months, no family history
- 1 patient after 36 months, history of angiofibroma scrotum resection, angiofibroma neck and shoulder, no family history

KAPLAN MEIER SURVIVAL FREE OF RECURRENCE CURVE



KAPLAN MEIER SURVIVAL CURVE



CONCLUSIONS

- Cardiac myxoma is a benign tumor with debilitating/lethal sequelae
- Urgent operation/low hospital mortality
- Low recurrence rate
- Survival of patients after myxoma removal does not significantly differ from the expected survival of the general population
- Genetic analysis/familial form
- Long-term follow-up with TTE