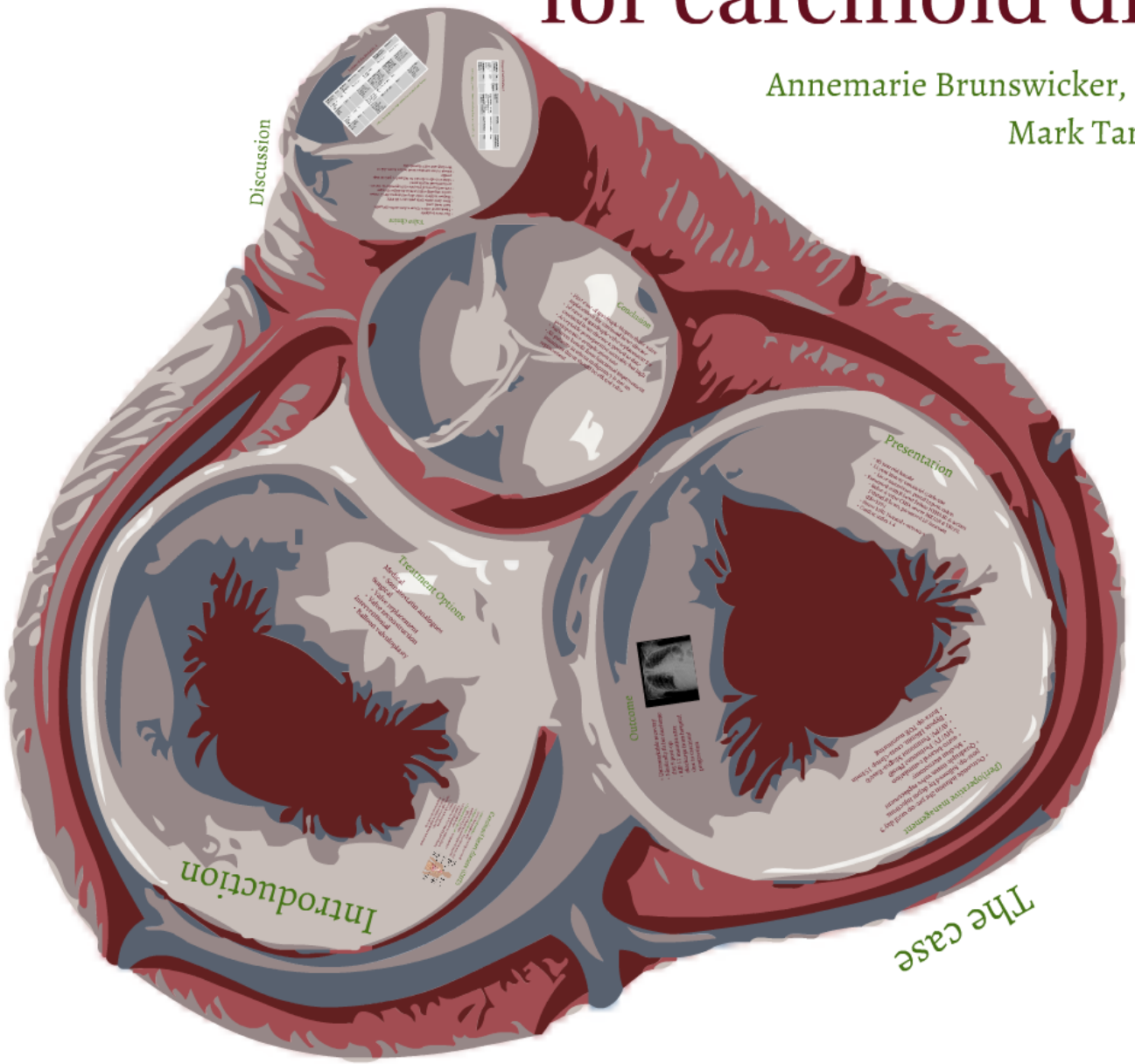
Papworth Hospital NHS
NHS Foundation Trust

Quadruple valve replacement for carcinoid disease

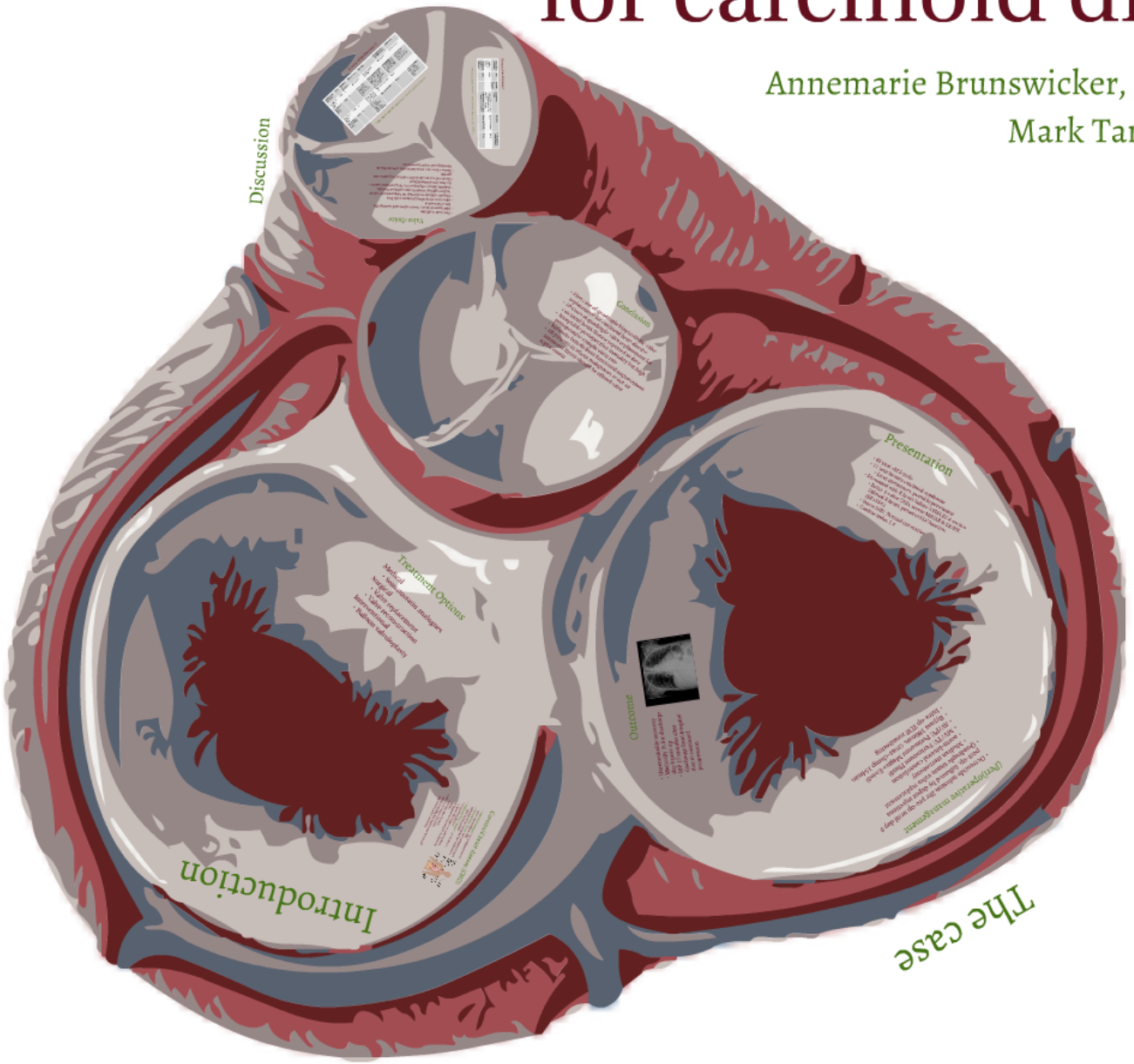
Annemarie Brunswicker, Marius Berman,
Mark Tan, John Dunning



Papworth Hospital NHS
NHS Foundation Trust

Quadruple valve replacement for carcinoid disease

Annemarie Brunswicker, Marius Berman,
Mark Tan, John Dunning



Introduction

Carcinoid heart disease (CHD)

- Carcinoid tumours: rare malignancies of enterochromaffine cells
- Carcinoid syndrome: liver metastases and release of vasoactive substances
- Carcinoid heart disease: secondary to systemic release
  - 60% develop CHD
- Carcinoid syndrome plaques on endocardium
  - carcinoid on tricuspid & pulmonary valves
  - Common lesions
  - 15% left sided for CHD
- Valve replacement but significant functional improvement
  - High mortality for survivors

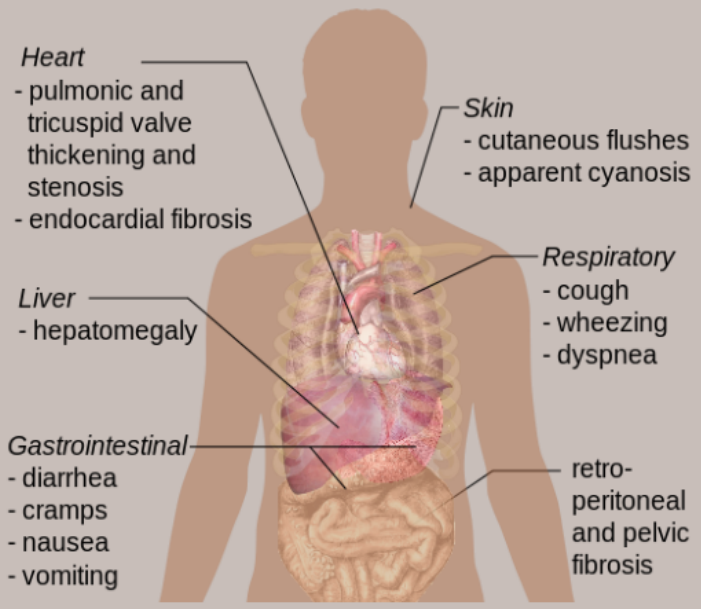
- Somatostatin analogues

Surgical
- Valve replacement
- Valve reconstruction

Interventional
- Balloon valvuloplasty

Carcinoid heart disease (CHD)

- Carcinoid tumours: rare malignancies of enterochromaffine cells
- Carcinoid syndrome: liver metastases and systemic release of vasoactive substances
  - 60% develop CHD
- Carcinoid heart disease: secondary to carcinoid syndrome
  - Carcinoid plaques on endocardium
  - Common on triscuspid & pulmonary valves
  - 15% left sided lesions
- Valve replacement for CHD
  - High mortality but significant functional improvement for survivors



Treatment Options

Medical

- Somatostatin analogues

Surgical

- Valve replacement
- Valve reconstruction

Interventional

- Balloon valvuloplasty

The case

(Peri)operative management

- Octreotide infusion 2hr pre-op until day 9 post-op, followed by depot injections
- Quadruple tissue valve replacement
- Median sternotomy
- aorto-bicaval cannulation
- MV/TV: Perimount Plus®
- AV/PV: Perimount Magna-Ease®
- Bypass 186min, cross-clamp 154min
- Intra-op TOE monitoring

- Unremarkable recovery
- Medically fit for discharge day 9 post-op
- RIP 17 months after discharge from hospital due to carcinoid progression

- history carcinoid syndrome
- Liver metastases, portal hypertension
- Presented with R heart failure NYHA III & ascites
- Echo: 4-valve CHD, severe MR/AR & TR/PR. Dilated R heart, preserved LV function (EF>55%)
- Stress MRI: Normal coronaries
- Cardiac index 1.4

Presentation

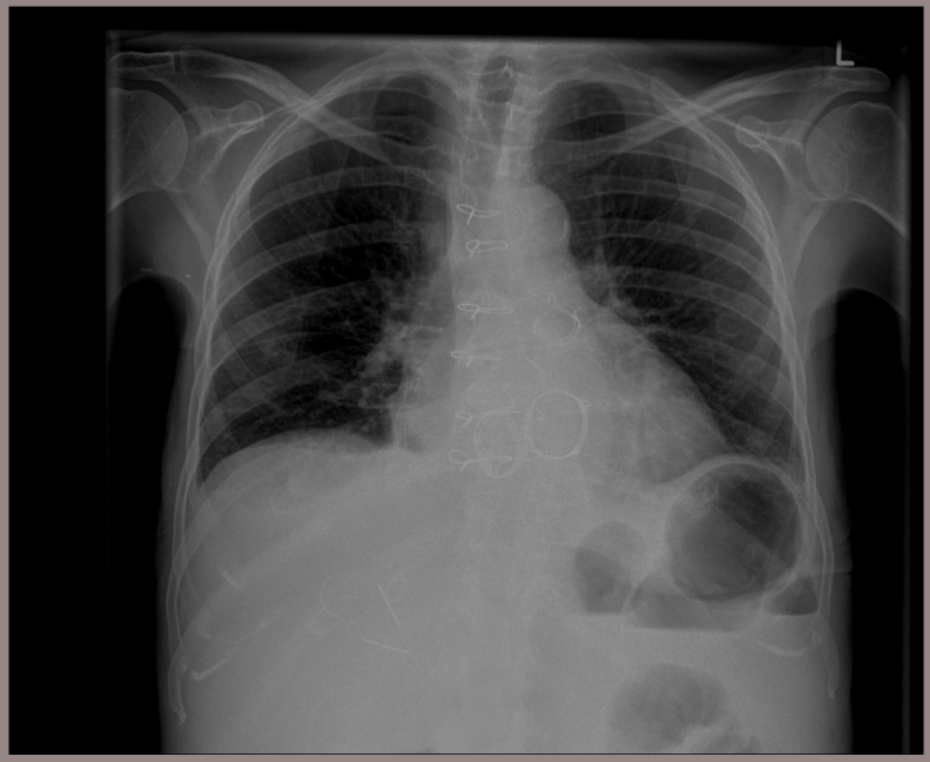
- 65 year old female
- 11 year history carcinoid syndrome
  - Liver metastases, portal hypertension
- Presented with R heart failure NYHA III & ascites
  - Echo: 4-valve CHD, severe MR/AR & TR/PR. Dilated R heart, preserved LV function (EF>55%)
  - Stress MRI: Normal coronaries
  - Cardiac index 1.4

(Peri)operative management

- Octreotide infusion 2hr pre-op until day 9 post-op, followed by depot injections
- Quadruple tissue valve replacement
  - Median sternotomy
  - aorto-bicaval cannulation
  - MV/TV: Perimount Plus®
  - AV/PV: Perimount Magna-Ease®
  - Bypass 186min, cross-clamp 154min
  - Intra-op TOE monitoring

Outcome

- Unremarkable recovery
- Medically fit for discharge day 9 post-op
- RIP 17 months after discharge from hospital due to carcinoid progression



Discussion

Review of the literature 1

Publication (Country)	Year	Number of patients	Valves used	Mortality	Postoperative complications	Comments
McAlindon et al [8] (UK)	2011	1	Not reported	No in-hospital mortality	Not reported	FO was closed
Arghami et al [9] (USA)	2010	7	St Jude (4), Carpentier-Edwards (1), Carbo-Medics (2)	1 in-hospital mortality secondary to heart failure, 3 deaths at 39, 5 and 30 months due to carcinoid progression	Atrial fibriliation (2), heart block (2), permanent pacing (1), AKI (1)	3 patients had FO closure. Octreotide used peri-operatively
Raja SG et al [10] (UK)	2009	1	Bioprosthesis (3), pulmonary homograft (1)	Alive at 1-year follow-up	None	
Knott-Craig CJ et al [11] (USA)	1992	1	St Jude Medical mechanical valve (3), pulmonary homograft (1)	Alive at 4 months	None.	30-day mortality in the entire cohort of 10 patients was 10%

Table 1: Quadruple valve replacements for carcinoid heart disease

Review of the literature 2

Publication (Country)	Year	Number of patients	Procedure/ Valves	Mortality	Postoperative complications
Castillo JG et al [12] (USA)	2008	1	Replacement of PV and TV, reconstruction of AV and MV, FO closure.	Alive at discharge	None
Chappin B et al [13] (Belgium)	2006	1	Replacement of PV and MV, PFO closure. TV plasty of AV and MV.	Alive at 25 months	None

Table 2: Valve replacements and reconstruction for quadruple valve carcinoid disease

Valve choice

- Few data available
- Mechanical valves, tissue valves and homografts have been used
- Most data stems from patients with TVR under ongoing somatostatin analogue therapy
- Plaques unlikely to develop on bioprosthetic valves
- Isolated reports of plaques on bioprosthetic valves - no functional impairment
- Choice of valves should be tailored to patient risk profile
- Tissue valves are associated with a lower risk of bleeding and valve thrombosis

Review of the literature 1

Publication (Country)	Year	Number of patients	Valves used	Mortality	Postoperative complications	Comments
McAlindon et al [8] (UK)	2011	1	Not reported	No in-hospital mortality	Not reported	FO was closed
Arghami et al [9] (USA)	2010	7	St Jude (4), Carpentier-Edwards (1), Carbo-Medics (2)	1 in-hospital mortality secondary to heart failure, 3 deaths at 39, 5 and 30 months due to carcinoid progression	Atrial fibrillation (2), heart block (2), permanent pacing (1), AKI (1)	3 patients had FO closure. Octreotide used peri-operatively
Raja SG et al [10] (UK)	2009	1	Bioprosthesis (3), pulmonary homograft (1)	Alive at 1-year follow-up	None	
Knott-Craig CJ et al [11] (USA)	1992	1	St Jude Medical mechanical valve (3), pulmonary homograft (1)	Alive at 4 months	None.	30-day mortality in the entire cohort of 10 patients was 10%

Table 1: Quadruple valve replacements for carcinoid heart disease

Review of the literature 2

Publication (Country)	Year	Number of patients	Procedure/ Valves	Mortality	Postoperative complications
Castillo JG et al [12] (USA)	2008	1	Replacement of PV and TV, reconstruction of AV and MV, FO closure.	Alive at 25 months	None
Chiappini B et al [13] (Belgium)	2006	1	Replacement of PV and TV, plasty of AV and MV.	Alive at discharge	None

Table 2: Valve replacements and reconstruction for quadruple valve carcinoid disease

Valve choice

- Few data available
- Mechanical valves, tissue valves and homografts have been used
- Most data stems from patients with TVR
- Plaques unlikely to develop on bioprosthetic valves under ongoing somatostatin analogue therapy
- Isolated reports of plaques on bioprosthetic valves - no functional impairment
- Choice of valves should be tailored to patient risk profile
- Tissue valves are associated with a lower risk of bleeding and valve thrombosis

Conclusion

- First case of quadruple bioprosthetic valve replacement for carcinoid heart disease
- 10 cases of quadruple valve replacement for carcinoid heart disease reported to date
- Acceptable postoperative mortality but high perioperative complication rate
- Survivors benefit from functional improvement
- All patients in whom malignancy is not an imminent threat should be offered valve replacement

Papworth Hospital NHS
NHS Foundation Trust

Quadruple valve replacement for carcinoid disease

Annemarie Brunswicker, Marius Berman,
Mark Tan, John Dunning