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P1**Long term follow-up of quadruple valve replacement for carcinoid heart disease**

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Introduction

Carcinoid heart disease most frequently involves the tricuspid or, more rarely, the pulmonary valve and presents with right heart failure as 5-HT is metabolized by the lung. Left-sided valve involvement is quite rare. We describe our experience of 3 patients presenting with heart failure secondary to carcinoid heart disease affecting all four cardiac valves. There are only four previous isolated case reports in the literature.

Methods

All three patients underwent quadruple valve replacement during a single operation. Right ventricular outflow tract reconstruction with a pericardial patch was performed in all patients. For 24 hours prior to surgery, all patients received intravenous Octreotide, which continued in intensive care for at least 24 hours.

Results

Mean cross-clamp and bypass times were 175 (range 164-197 minutes) and 210 (range 195-229 minutes) minutes, respectively. Mean intensive treatment unit (ITU) and inpatient stays were 2.3 (range 2-3 days) and 12 (range 9-16 days) days, respectively. One patient was reopened for bleeding 4 hours postoperatively from a ventricular pacing wire site. None required a permanent pacemaker post-operatively. There were no other complications in any patient. The quality of life was excellent at 58-70 months clinic follow-up as they were in NYHA 1-2. Postoperative echocardiography showed no para-valvular leaks and well-functioning prostheses in all cases.

Conclusion

Surgery to replace all four valves is feasible with excellent medium-term survival and a very low rate of complications. Patients with carcinoid heart disease should always be considered for surgery irrespective of the extent of valvular involvement.

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P2**Carcinoid heart disease: Early outcomes after surgical valve replacement in thirteen patients**

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Aim

To describe the early outcomes of carcinoid patients undergoing surgical heart valve replacement.

Methods

In a retrospective study, records of patients with symptomatic carcinoid heart disease referred for valve surgery between 2012 and 2021 were reviewed. The perioperative and early postoperative outcomes were analyzed.

Results

Thirteen patients, with a mean age of 64 years (range 55-79 years) underwent cardiac surgery for carcinoid syndrome. 3 patients had quadruple valve replacement, 7 had tricuspid and pulmonary valves changed, 2 had tricuspid valve replacement, while one had tricuspid, pulmonary, and aortic valves replaced. Right-sided valves were replaced with biological valves in 12 patients and a mechanical valve in 1 patient. Left-sided valves were replaced with a mechanical valve in 2 patients and with a biological valve in 1 patient. Mean postoperative follow-up was 56 months (range 2-102 months, median 65 months). All patients had a good left ventricle except one, in whom it was mildly impaired. The right ventricle was severely dilated in 4 patients, moderately in 4, and mildly in 3. One patient died of heart failure 10 days postoperatively and one patient succumbed to Acute Carcinoid Crisis 8 days after surgery. Functional improvement was noted in all survivors, and they were all in New York Heart Association class I-2 at last follow up and none required permanent pacemaker. One patient capitulated to neuro-endocrine tumour at nearly 6 years, giving us survival rate of 91% at 56 months (range 2-102 months). Nobody required any valve intervention in the follow-up period.

Conclusion

Although carcinoid syndrome is a rare and progressive disease, valve replacement in symptomatic patients is a reasonable option with survival benefit, low early postoperative mortality, without valve-related complications, and with functional improvement. Cardiac assessment is required in all patients with carcinoid disease to ensure that appropriate patients are put forward for surgery when symptomatic.

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P3**Enhanced expression of the proinflammatory, profibrotic molecule, vascular adhesion protein-1 in carcinoid heart disease valves**

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Title

Enhanced Expression of the Proinflammatory, Profibrotic Molecule, Vascular Adhesion Protein-1 in Carcinoid Heart Disease Valves

Introduction

Vascular adhesion protein-1 (VAP-1) is a known driver of hepatic inflammation and fibrosis, with elevated levels detected in the circulation in disease. We studied the tissue expression of VAP-1 in midgut NETs and carcinoid heart disease (CHD) alongside circulating soluble VAP-1 (sVAP-1) to determine the role of VAP-1 in NETs.

Methods

Expression of VAP-1 and inflammatory and stromal markers in midgut NETs tissue, CHD valves and control valves was assessed using immunohistochemistry. sVAP-1 concentration was measured by ELISA in serum collected from two independent NET centres.

Results

In midgut NETs VAP-1 protein expression was predominantly localised to fibroblast-rich areas. Tissue expression of VAP-1 was markedly increased in CHD valves ($n=33$) compared to control valves ($n=6$, aortic/mitral degenerative disease; $P>0.001$) and localised to neovessels and the plaque. VAP-1 expression increased with plaque maturity with myxoid regions greater than collagen and elastin rich areas. Significantly higher levels of sVAP-1 were found for midgut NETs (median 1005 ng/ml, IQR 790-1148, $n=50$) compared with controls (median 740 ng/ml, IQR 535-867, $n=12$; healthy volunteers; $P=0.0014$). Soluble VAP-1 levels remained high in patients with CHD (median 1280 ng/ml, IQR: 961.2-1652.0; $P>0.001$ vs controls, $n=17$). We confirmed an increase of sVAP-1 in an independent cohort (median 661 ng/ml, IQR 514-883 vs median 285 ng/ml, IQR 211-480; $P>0.001$), for midgut NET patients ($n=40$) vs controls ($n=10$; benign gynaecological disease).

Conclusions

VAP-1 protein expression is associated with fibrotic disease in both midgut NETs and CHD, with concomitant increases in circulating sVAP-1. These studies implicate VAP-1 in the pathophysiology of midgut NETs and CHD, with potential utility as a stratification tool and novel therapeutic target.

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P4**The real value of the NET CNS led specialist carcinoid heart disease service: Making it happen**

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Introduction

The diagnosis of carcinoid heart disease (CHD) is often made late and the patients are often deconditioned, with symptoms of fatigue, weight loss and refractory carcinoid symptoms. The challenge is then to improve the patient's condition by an enhanced pathway of prehabilitation and interventions, in order to consider surgical and medical therapies that can improve the patient's quality of life and prognosis. In 2018 due to the complexity of patients and increased referrals to our CHD service, it became unmanageable to look after these patients through the current ad hoc service provision and so we needed to build closer collaborations with our cardiac colleagues. It is recognised that the health care resources to manage CHD are not well understood. The Neuroendocrine Tumour (NET) Clinical Nurse Specialist (CNS) team recognised the unmet need for this complex group of patients and were instrumental in bringing together the wider members of the specialist teams. This culminated in a twice monthly CHD/NET multidisciplinary team meeting (MDTM) and a monthly joint NET/CHD clinic.

Methods

Patient information was collected on a prospective database designed and maintained by the CNSs in 2018, in addition to the information on the Trust NET database. This study compared the patient data collected on a standard pathway from 2005, prior to CHD/NET service developments in 2018, to those on the enhanced pathway from December 2018 to present.

Results

Results show that 45 patients with CHD were reviewed at NET CHD MDT from December 2018 to July 2021. Nineteen patients went on to have had heart valve replacement and of these four had patent foramen ovale (PFO) closure prior to proceeding to surgery. A number of patients benefitted from clinical review and optimisation in the innovative joint CHD /NET clinic.

Conclusions

Establishing a dedicated CHD MDT and joint CHD/NET clinic was fundamental in driving this service forward to improve efficiencies in the CHD service and overall patient care. There is growing evidence that the CNS role makes a difference to patient care, however the full value is not always recognised. Further analysis could lead the way for future developments and improvements in the service.

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P5

Nutritional recovery is essential for successful management of patients with advanced carcinoid heart disease

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Introduction

Chronic heart failure is a catabolic state leading to cachexia in 5-15% of patients, predicting poor survival outcomes. Malnutrition affects 40% of patients with NETs and 58% with carcinoid syndrome (CS).

Case

45M with severe carcinoid heart disease, on referral and admission was cachectic with BMI of 16, having lost 32 kg or 30% of body weight, meeting 50% of dietary requirements and at risk of refeeding syndrome. Nasogastric feeding (NG) was

started with 1L Nutrison energy Multifibre. Six days later he was also managing all 3 meals, snacks, snack box, and cooked breakfast daily. Fortisip compact protein BD provided additional 600kcal and 36g protein. Steatorrhea occurred due to octreotide. Creon 25,000U 2-3 with meals and 1-2 with snacks/ ONS prescribed, titrated up 27/3/2019 for persistent steatorrhea. Nutritional requirements were calculated based on 52 kg weight at 1820kcal (35kcal/kg) + 65g protein (0.2g of nitrogen). NG feed changed to peptide based peptisorb 1.8L providing 1800kcal and 72g protein daily from feed alone. Urinary 5HIAA was 1189 on admission; suffering with severe CS and mini carcinoid crises on standing despite receiving sandostatin LAR 30mg 4 weekly. Considerably short of breath at rest, NYHA class 4 with orthopnoea and PND. CS was managed with increase in sandostatin 30mg frequency to 2 weekly, addition of octreotide 100mcg s/c tds and telotristat 250mg p.o. tds. 5HIAA improved to 429 before cardiac interventions. Weight plateaued by the 19/4/2019; NG feed was increased. 12/6/2019 he weighed 67.4 kg BMI-22 kg/m² (+17.7 kg since admission). PFO closure completed 21/6/2019. Weight 70.6 kg 28/6/2019. He was functionally improved, feeling fitter and stronger, using an exercise bike for 5 mins per day - 1 mile. AR/PR/TR valve surgery 12/7/2019 and continued on NG feed.

Outcomes

24/07/2019 NGT out. He continued with fortisip compact protein TDS as an additional top up providing a further 900kcal and 54g protein. Weight 74.1 kg at discharge and reported visually stable over following 8 months. 5HIAA 35 (normal) at discharge. From mid-admission 02/05/19 to discharge on 7/8/2019: Minnesota living with heart failure score improved from 83 to 19; EORTC QLQC30 from 88 to 56 and GINET21 from 50 to 34.

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