

Carcinoid Heart Disease: Treatment Challenges

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ABSTRACT

Carcinoid Heart Disease (CHD) or Hedinger syndrome is a serious complication of carcinoid syndrome or what is called Neuroendocrine Tumor (NET), it may occur in up to 60% of patients with carcinoid syndrome or can be the initial presentation in up to 20% and it is associated with poor survival. It usually affects the right side of the heart, especially, the tricuspid and pulmonic valves, and leads to valvular regurgitation and lesser extent stenosis due to inflammation and plaque deposits caused by the circulating serotonin and other vasoactive peptides secreted by NET. Small intestine and lung are the two most common sites of NET and small intestine is the commonest primary site. CHD treatment is challenging and it could be either medical early in the course of the disease but may need surgical intervention with valve replacement in the advanced disease with right heart failure.

Keywords: Carcinoid heart disease; Carcinoid syndrome; Neuroendocrine tumor; Tricuspid valve; Pulmonic valve

DESCRIPTION

N-Terminal Pro-brain Natriuretic Peptide (NT ProBNP) is an important serum screening and surveillance marker for CHD in patients with carcinoid syndrome and transthoracic echocardiography remains the gold standard diagnostic modality for patients with NT ProBNP > 260 pg/ml or in symptomatic individuals, but for the asymptomatic patients with NT ProBNP < 260, 6-months surveillance by checking NT ProBNP and clinical assessment is required [1-3]. Other biomarkers used for screening and surveillance are Chromogranin-A (CgA), and urinary 5-Hydroxy-Indoleacetic Acid (5-HIAA), a metabolite of 5-Hydroxytryptamine (5-HT) [4]. Treatment of CHD may be medical and surgical depending on the right heart failure status, the severity of the valve disease, and how extensive is the neuroendocrine disease [5]. Medical treatment is to control the symptoms but neither slow the progression of the disease nor improve survival, somatostatin analogs like octreotide, lanreotide, and pasireotide are used as the main medical treatment and they act by slowing the release of serotonin into the circulation [5]. Some other combined chemotherapeutic agents are also used to treat NET like combined trastuzumab, paclitaxel, and interleukin 12 for NET with epidermal growth factor 2/neu expression, and telotristat used to treat carcinoid-

induced diarrhea by inhibiting serotonin release [5]. The mainstay surgical treatment is valve replacement and it improves survival and quality of life [5].

Valve replacement is indicated in symptomatic patients with right ventricular heart failure and severe pulmonic or tricuspid valve disease and expected survival ≥ 1 year, however, valve replacement should also be considered for asymptomatic patients with severe pulmonic or tricuspid valve disease and expected survival ≥ 1 year [3,5]. Contraindications to surgical intervention are the advanced metastatic NET and poorly controlled NET while on octreotide therapy or even after trans-arterial embolization of hepatic carcinoid metastasis, bioprosthetic valves are preferred over mechanical valves due to the lack of long-term anticoagulation especially with the coagulopathy caused by the extensive carcinoid liver metastases [5]. Prosthetic valves require 3-6 months of anticoagulation after the valve's implantation [5]. Studies showed that valve replacement is linked with better symptom control and longer survival in comparison with medical treatment (40% at 2 years vs 8%).⁴ Percutaneous transcatheter valve implantation is an emerging less invasive novel approach, especially for those with high comorbidities and who can not tolerate open heart surgery, also it is a reasonable approach for those who require redo surgery for incompetent bioprosthetic valves with valve-in-valve approach [6,7].

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CONCLUSION

CHD is a serious life-threatening complication of NET associated with poor quality of life and shorter survival even with aggressive medical treatment especially the extensive NET metastases. Valves replacement is indicated in symptomatic right heart failure, and it improves survival and quality of life. Percutaneous transcatheter is a newly emerging alternative option with better tolerance for the highly morbid patients and for those with degenerative bioprosthetic valves.

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