

REVIEW

A Review of Open and Endovascular Treatment of Superior Vena Cava Syndrome of Benign Aetiology

G.S. Sfyroeras ^{a,*}, C.N. Antonopoulos ^a, G. Mantas ^a, K.G. Moulakakis ^a, J.D. Kakisis ^a, E. Brountzos ^b, C.R. Lattimer ^c, G. Geroulakos ^{a,c}

^a Department of Vascular Surgery, National and Kapodistrian University of Athens Medical School, Attikon University Hospital, Athens, Greece

^b 2nd Laboratory of Radiology, National and Kapodistrian University of Athens Medical School, Attikon University Hospital, Athens, Greece

^c Department of Surgery and Cancer, Imperial College, London, UK

WHAT THIS PAPER ADDS

There is an increased incidence of benign causes of superior vena cava syndrome (SVCS) mainly as a result of the use of intravenous devices such as central venous catheters, pacemakers, and defibrillators. This review of the indications, technical details, and the results of open and endovascular treatment of benign SVCS shows that while surgery is used for more advanced cases of SVCS, both techniques appear to be effective in relieving clinical symptoms and there is a need for continuous follow-up and re-intervention to achieve good early and mid-term results.

Background: The widespread use of central venous catheters, ports, pacemakers, and defibrillators has increased the incidence of benign superior vena cava syndrome (SVCS). This study aimed at reviewing the results of open and endovascular treatment of SVCS.

Method: Medical literature databases were searched for relevant studies. Studies with more than five adult patients, reporting separate results for the SVC were included. Nine studies reported the results of endovascular treatment of SVCS including 136 patients followed up for a mean of 11–48 months. Causes of SVCS were central venous catheters and pacemakers (80.6%), mediastinal fibrosis (13.7%), and other (5.6%). Percutaneous transluminal angioplasty (PTA) and stenting was performed in 73.6%, PTA only in 17.3%, and thrombolysis, PTA, and stenting in 9%. Four studies reported the results of open repair of SVCS including 87 patients followed up between 30 months and 10.9 years. The causes were mediastinal fibrosis (58.4%), catheters and pacemakers (28.5%), and other (13%). Operations performed included a spiral saphenous interposition graft, other vein graft, PTFE graft, and human allograft. Thirteen patients required re-operations (15%) before discharge mainly for graft thrombosis.

Results: In the endovascular group technical success was 95.6%. Thirty day mortality was 0%. Regression of symptoms was reported in 97.3%. Thirty-two patients (26.9%) underwent 58 secondary procedures. In the open group the 30 day mortality was 0%. Symptom regression was reported in 93.5%. Twenty-four patients (28.4%) underwent a total of 33 secondary procedures.

Conclusions: Endovascular is the first line treatment for SVCS caused by intravenous devices, whereas surgery is most often performed for mediastinal fibrosis. Both treatments show good results regarding regression of the symptoms and mid-term primary patency, with a significant incidence of secondary interventions.

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INTRODUCTION

Superior vena cava syndrome (SVCS) is the collection of clinical symptoms and signs resulting from obstruction of blood flow through the superior vena cava (SVC). While many cases are caused by malignant lesions of the mediastinum, some are related to benign causes, including mediastinal fibrosis, post-radiation therapy and secondary to placement of central venous catheters, pacemakers, and

* Corresponding author. Department of Vascular Surgery, "Attikon" University Hospital, 1 Rimini Street, 124 62, Athens, Greece.

E-mail address: gsfyr@yahoo.gr (G.S. Sfyroeras).

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defibrillators.¹ Approximately 19,000 cases of SVCS occur every year in the USA, with increasing frequency concomitant to the increased use of intravenous devices.² Benign aetiologies may now comprise up to 40% of cases.³

Symptoms of the syndrome vary mainly according to the speed of onset of the obstruction to flow. When obstruction develops slowly and progressively, a collateral circulation develops and symptoms are mild or absent. However, rapid obstruction can lead to oedema of the face, neck and arms, dyspnoea, cerebral oedema, and cerebellar herniation.⁴

There are four types of SVCS (I-IV) according to Stanford and Doty.⁵ In type I there is high grade SVC stenosis but a normal direction of blood flow through the SVC and azygos veins. There is also an increased collateral circulation through the hemiazygos and accessory hemiazygos veins. In type II there is a >90% stenosis or occlusion of the SVC. This is also associated with a normal direction of blood flow through the azygos vein. In type III there is occlusion of the SVC with retrograde flow in both the azygos and hemiazygos veins. In type IV there is extensive occlusion of the SVC, innominate, and azygos veins with chest wall and epigastric venous collaterals.⁵ The aim of this study was to review the results of open and endovascular treatment of SVCS of benign aetiology.

MATERIALS AND METHODS

Data collection

The current study followed the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines.⁶ Medline, Scopus, EMBASE, Google Scholar, Ovid, and the Cochrane Library medical literature databases were searched. After retrieving the relevant articles, their individual reference lists were evaluated to retrieve additional articles. These were included in the analysis provided the inclusion criteria were satisfied. Risk of bias in the eligible studies was assessed by each reviewer using the Newcastle-Ottawa Quality Assessment Scale.

Search methodology and data extraction

The medical subject headings (MESH) terms used were: ("endovascular"[All Fields] OR "open"[All Fields]) AND "vena cava"[All Fields] AND ("thrombosis"[All Fields] OR "occlusion"[All Fields]). Studies published in English investigating endovascular or open repair for benign SVC syndrome were searched for. Studies including more than five adult patients, reporting separate results for the SVC were included. The last search was performed in March 2016. Studies which did not report treatment outcomes for SVCS, studies which reported outcomes for malignant SVCS, and studies which did not report separate outcomes for SVC because other brachiocephalic veins were also evaluated, were excluded. Other exclusions were case series with fewer than five cases, and publications on overlapping populations. An additional secondary literature search was performed in PubMed Advanced Search Builder with Query: Vena Cava AND thrombosis AND treatment NOT medical.

Literature search, study selection, data extraction, and data analysis were independently performed by two authors (CNA, GM), and the final decision was reached by consensus. Data extracted from eligible studies included the first author's name, study year, country in which the study was conducted, type of study, study period, total number of patients, indications for treatment, aetiology of SVC syndrome, risk factors for SVCS, radiological imaging performed, SVCS classification, operation performed, graft material used for open repair, follow-up (years), primary patency, assisted primary patency, secondary patency rates, 30 day mortality, re-operations during follow-up, complications, clinical success, endovascular technique used (primary PTA or PTA + stent), mean age (years), males (%), and type of balloons and stents used.

Statistical analysis

Data synthesis and treatment effects. Primary, assisted primary, and secondary patency rates were extracted from the eligible studies. If the rates were not reported, they were calculated from the study by recording the number of patients with patent SVCs together with the total number of patients at a given time point. If there were no numerical values reported in the text, patency rates were extrapolated from the Kaplan-Meier curves using the automatic tools of line drawing and distance measure provided by GetData Graph Digitizer v2.26 (www.getdata-graph-digitizer.com). Values of the respective patency rates were subsequently calculated and expressed as proportions with 95% CIs. These were later transformed into quantities according to the Freeman-Tukey variant of the arcsine square root transformed proportion. The pooled effect estimates were calculated as the back-transformation of the weighted mean of the transformed proportions, using inverse arcsine variance weights for the fixed effects model and DerSimonian-Laird weights for the random effects model.⁷ The random effects model was necessary because the differences between the studies were often large and very inconsistent. Pooled patency rates of the endovascular and open approaches were reported at 1, 2, 4, 8, 12, 26, and 36 months.

Heterogeneity and publication bias. A formal statistical test for measuring heterogeneity was performed using the I^2 test. Publication bias was assessed using the Begg-Mazumdar adjusted rank correlation, as well as from a visual inspection of the funnel plots. The statistics were conducted using Stata v13.1 (College Station, TX, USA).

RESULTS

Study characteristics

As shown on the PRISMA flow chart (Fig. 1), a total of 575 articles were identified in the review for consideration. With the endovascular approach, nine studies (136 patients) were finally deemed eligible after applying the inclusion/exclusion criteria.^{4,8–15} For open surgical repair, four studies (87 patients) were finally eligible.^{10,16–18} All

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