Audit

Current management of Paget-Schroetter syndrome in the UK

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Introduction: Untreated symptomatic patients with Paget-Schroetter syndrome (PSS) can sustain chronic disability from venous obstruction, with arm swelling, pain and early exercise fatigue. This may result in significant loss of occupational productivity and quality of life. For this reason, active management is recommended in the majority of the recent literature. The objective of this study was to assess current trends of management of PSS in the UK.

Methods: A 9-part questionnaire was sent to 90 ordinary members of the Vascular Surgical Society of Great Britain and Ireland (VSS-GBI). Names and addresses were selected by highlighting every fourth ordinary member from the UK, in the 2000 VSS-GBI handbook. Ordinary members of the VSS-GBI who were clearly radiologists were excluded.

Results: Of the 90 questionnaires sent, 60 were returned (66.67%). The majority of respondents used both duplex and venography (61%) as the major investigative tools though some employed duplex only (17%). Multimodality treatment (radiological and operative) was the favoured approach. Only 17% still favoured conservative management alone. Thrombolysis was the most common intervention (86.7%) usually followed by elective thoracic outlet decompression. Most favoured a delayed approach for surgery of 6–12 weeks. First rib resection was the most commonly performed operation (58%), usually by the transaxillary approach (55%). Most of the respondents were doubtful of the role of stenting in this condition and did not use it.

Conclusions: There is no definite consensus on treatment of this condition in the UK. A majority tend to favour a multimodal approach. Thrombolysis is the most common form of treatment employed and first rib resection via the transaxillary approach remains the most popular surgical procedure. The lack of consensus of this potentially disabling condition highlights the need for randomised clinical trials to guide management.

Key words: Primary subclavian vein thrombosis – Management

Paget-Schroetter syndrome (PSS; primary subclavian-axillary vein thrombosis) is potentially a disabling condition. Untreated, it can lead to upper limb swelling, pain and exercise fatigue.^{1–5} This not only leads to decreased productivity but also to reduced quality of life.⁶

The pathophysiology of the condition is little understood and this has resulted in varied and diverse management.^{6–10} Recent literature, mainly from North America, predominantly recommends an active management protocol. However, it is clear from discussion with colleagues that

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conservative management with simple anticoagulation is still widely used in the UK and Europe. In addition, there seems to be no consensus in the literature with regard to the timing and precise nature of active management and there is presently no agreed protocol for the optimum management of PSS in the UK. Controversy exists partly because no randomised controlled studies exist. We aimed to assess the current approach to management of Paget-Schroetter syndrome in the UK by means of a postal questionnaire.

Methods

A questionnaire (Appendix) was sent out to 90 ordinary members of the Vascular Surgical Society of Great Britain and Ireland (VSS-GBI). Names and addresses were selected by highlighting every fourth ordinary member from the UK, in the 2001 VSS-GBI handbook. Ordinary members of the VSS-GBI who were clearly radiologists were excluded.

The 9-part questionnaire was sent out with a stamped-addressed envelope to the selected individuals. It was designed in keeping with the multiple approaches detailed in the current literature. Questions 1–3 detailed the status of the surgeon and number of cases of PSS seen. Question 4 was related to use of various treatment modalities. Questions 6–9 were related to the surgical management of the condition. The questionnaire was initially tested within our department and any pitfalls were corrected before its circulation to the participants. The questionnaire was intentionally limited to one side of two A4 sheets in an attempt to improve the response rate. The questionnaires were sent out in late February 2002 and the closing date for responses was 12 weeks later. Responses were anonymised and hence no reminders were sent.

Results

Of the 90 questionnaires sent, 60 were returned (66.67%). Of the 60 respondents, 39 were vascular surgeons and 21 general surgeons with a vascular interest. Four of the surgeons did not offer their comments, while 5 of the surgeons who had not seen any case in last 2 years did comment on their management options. Only 9 (15%) had seen no case of PSS in the last 2 years, 31 (51.6%) had seen three or more cases during this period, while 16 (26.6%) had seen five or more.

Investigation methods

Duplex scanning along with venography (61%) were the preferred investigations, although some surgeons used

Figure 1 Initial investigation(s) in Paget-Schroetter syndrome.

Figure 2 Treatment patterns in Paget-Schroetter syndrome.

duplex alone (17%). Magnetic resonance venography was also employed by few in conjunction with duplex and venogram or duplex alone (Fig. 1).

Treatment options

Multimodality treatment (radiological and operative) was the most commonly favoured approach. Only 17% of the surgeons still favoured conservative management alone (Fig. 2). Thrombolysis was the most common intervention (86.7%) among surgeons favouring more active management. This was usually followed by elective thoracic outlet decompression. Around two-thirds of the respondents would employ surgery if desired (Fig. 3). Less than half would employ angioplasty at some stage, mainly following thrombolysis. Only one surgeon would employ stenting as the primary treatment. One-third would use it as an adjunct to other modalities while the majority of respondents (65%) were either doubtful or did not consider there was a role for stenting in this condition.

Figure 3 Choice of interventions.

Figure 4 Timing of surgery following diagnosis.

Procedure of choice

A majority of surgeons would opt for surgical treatment (64% versus 36%; Fig. 4). A quarter of these offered surgical treatment as the primary treatment. Of the five options offered in the questionnaire, first rib resection alone was the preferred procedure (74%; Table 1). A few respondents selected more than one option to represent their usual practice.

Surgical approach

A transaxillary approach was more popular than supraclavicular (55% versus 28%). Some preferred a combination of the given options (Table 2).

Timing of the surgery

Most favoured a delayed approach for surgery at an interval of 6–12 weeks though 21% of the surgeons commenting on the option, would go for early surgery within 48 h of the diagnosis (Fig. 4). Some commented on the importance of considering the presence of any residual symptoms and thus their timing would be variable.

Discussion

Population-based studies have estimated the age-adjusted incidence of deep vein thrombosis at 48 per 100,000 persons per year, increasing steadily with age.^{11,16} Of all cases of deep venous thrombosis, thrombosis of the upper extremity veins comprises only in the order of 1–4% and primary subclavian or axillary vein thrombosis accounts for approximately 20% of these patients.^{1,11–18} Although unusual, it is not exceptionally rare. In the UK, more than quarter of the surgeons in our survey had managed five or more cases in the last 2 years.

Table 1 Preferred surgical procedure

Surgical procedure	n (%)
First-rib resection	35 (74%)
Scalenotomy	1 (2%)
Rib resection + scalenotomy	3 (6%)
Rib resection + scalenectomy	5 (10%)
Variable	3 (6%)

Table 2 Preferred surgical approach among surgeons favouring surgery

Surgical approach	n (%)
Axillary	26 (55%)
Supraclavicular	13 (28%)
Paraclavicular	2 (4%)
Axillary + supraclavicular	6 (13%)

Duplex ultrasonography has established itself as the primary imaging technique in suspected cases of PSS.^{19,20} It provides a safe, reliable, and repeatable method of evaluating and following patients.^{19,21} Colour Doppler and measurements of velocity change have helped further to improve the sensitivity of the sonographic techniques. Longley *et al.*¹⁸ showed that when the sonographic criteria of subclavian vein clot or compression resulting in a complete loss of normal venous phasicity with arm abduction and the clinical criterion of subsequent improvement in symptoms after curative surgery are used, colour Doppler sonography is 92% sensitive and 95% specific for the diagnosis of PSS.

Contrast venography is also commonly used for the diagnosis of subclavian vein thrombosis as it allows the direct visualisation of the whole deep-vein system of the arm and also allows continuation to recanalisation of local thrombolysis. Digital subtraction has not only allowed

more precise images but also lower doses of contrast material to obtain the appropriate images.²³ Positional or provocative venography, by showing any costoclavicular compression of the subclavian/axillary vein on abduction and external rotation of the upper extremity, may also help in identifying those patients who may be at risk of recurrent thrombosis.^{24,25} Lee *et al.*²⁶ have emphasised the importance of looking for evidence of compression of the collateral vessels, as well as the main vein with such provocative manoeuvres. This helps to determine not only the haemodynamic significance but also the chronicity of the occlusion. A majority of surgeons in our survey employed combined duplex and venography, a few used duplex alone, and others added magnetic resonance venography or a spiral computed tomography.

Symptomatic patients treated conservatively may sustain chronic disability from venous obstruction, with arm swelling, pain, and early exercise fatigue.^{2-4,27} A number of patients may also develop more extensive symptoms of neurogenic thoracic outlet syndrome.^{5,28,29} A history of antecedent strenuous or repetitive activities is present in more than 75% of the patients and in around 70% the condition affects their dominant side.^{1,2,4,5,30-33} As the condition usually affects younger patients, there may be serious implications for subsequent employment if symptoms persist. Such persistent sequelae and disability has been observed in 20-75% of patients in long-term follow-up series.^{1-5,35-37} This observation has prompted a shift towards more active treatment in the literature. However, there has never been a randomised comparison between active and conservative treatment for PSS. Of our respondents, 17% still prefer a conservative approach in favour of anticoagulation alone. Thrombolysis is mainly applied in the acute setting and can produce recanalisation in 90% of the cases when employed within the first week of thrombosis.^{15,38-40} In our survey, more than 86% of the surgeons who would employ intervention in PSS used thrombolysis initially.

The published literature suggests that balloon angioplasty has little to offer in patients with pre-operative dynamic compression of the subclavian or axillary vein in the costoclavicular space and is unlikely to improve luminal calibre without thoracic decompression.^{20,22,31,42} However, percutaneous angioplasty may be useful in cases of residual stenosis where the decompressive surgery has already been performed.⁴¹ Re-thrombosis or restenoses are likely to occur without thoracic decompression.^{40,43-45} Our survey suggests that, in the UK, angioplasty of the subclavian vein is still used but usually only as a part of a multimodal approach rather than for primary treatment.

Primary stenting also does not appear to have many supporters in the UK. Around half of the respondents do

not see a role or would not employ stenting in PSS while others would use it only as an adjunct to other modalities. Clearly, lesions that are seen on completion venography after successful thrombolysis usually represent extrinsic compression from fixed, musculoskeletal structures of the thoracic outlet and cannot be corrected with adjunctive stenting. Often, the result is deformation or fracture of the stent, occlusion and at times injury to the vein.^{24,26,46-48} Because of the high incidence of complications associated with stents, their role without thoracic decompression is very limited. They may occasionally be useful in cases of post-surgical decompression of intrinsic stenotic venous lesions.

Simple division of the scalene and subclavius muscles or the surrounding fibrous structures may not be enough to decompress the thoracic outlet adequately.²⁴ First-rib resection was clearly the most popular surgical procedure among the UK surgeons; among our respondents favouring surgery, 74% will do the first-rib resection alone while another 16% will combine this with scalenotomy or scalenectomy. The axillary approach is preferred as it gives relatively good exposure of the anterior end of the first rib compared with the supraclavicular route. This limits any damage on the collaterals and allows adequate subclavian vein exposure and venolysis. It may, however, provide limited exposure for more extensive procedures. In the UK, more than half of the surgeons (55%) preferred the axillary approach as compared to supraclavicular (28%) and 13% would employ a combined axillary and supraclavicular approach.

There is considerable debate in the published literature about the optimum timing for intervention. Some advocate expeditious thrombolytic therapy and early surgery (first-rib resection).⁴⁹⁻⁵¹ Others have advocated early thrombotic therapy and an interval period of anticoagulation, followed by late (4 weeks to 6 months) surgical decompression of the thoracic outlet.^{31,52-53} In our survey, 25% of the surgeons who tend to operate, will do so within 48 h. However, a majority still favour a 6-12 week delay. One rationale behind this may be to allow a reduction and clearing of the acute inflammatory process prior to surgery and, to a certain extent, to allow the selection of appropriate cases for surgery based on residual symptoms. In the absence of randomised studies, both approaches can be considered acceptable based on current literature.

Rutherford and Hurlbert⁵⁴ administered a multiple-choice questionnaire to 25 US vascular surgeons with a special interest in subclavian vein thrombosis. The majority agreed upon recommending the following principles in the management of PSS: (i) early removal of clot in active, healthy patients; (ii) catheter-directed thrombolysis as initial therapy; (iii) further therapy based on follow-up positional venography; (iv) surgical relief of

demonstrated thoracic outlet compression after a brief period of anticoagulation therapy; (v) conservative therapy if post-lysis venography showed either no extrinsic compression or a short residual occlusion; and (vi) intervention for residual intrinsic lesions with over 50% narrowing.

Conclusions

This survey has demonstrated that there is lack of consensus on the optimal management of Paget-Schroetter syndrome in the UK. The majority of vascular surgeons prefer to use a multimodal approach. Thrombolysis remains the most popular choice followed closely by decompression surgery of the thoracic outlet. Anticoagulation alone as a primary treatment is losing favour. The study has also demonstrated that the most popular operative management option is first-rib resection via the axillary approach, with 6–12 weeks being the optimal time favoured by most for thoracic decompression. The use of stenting in PSS remains dubious. In view of such diverse possible treatment options, there is need for a comparison of different modalities in randomised, prospective trials.

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APPENDIX

QUESTIONNAIRE on Paget-Schroetter syndrome (primary subclavian-axillary vein thrombosis)

1. Your status?
2. How many cases of primary subclavian vein thromboses have been under your care (or joint care with another colleague) in the last two years?
3. What are your diagnostic tools besides clinical examination?
4. Which of the following treatment modalities do you usually employ?
5. If you use stenting of the subclavian/axillary vein, is it:
 - (1) A primary treatment?
 - (2) As an adjunct to thrombolysis?
 - (3) As an adjunct to surgery?
 - (4) As an adjunct to angioplasty?
 - (5) No role to play?
6. If you use surgery of the thoracic outlet, what are your procedures of choice?
7. If first-rib resection, which approach do you use?
8. What timing of surgery do you favour?
9. If late decompression, how long do you wait?