

The Paget-Schroetter Syndrome: Work Accident and Occupational Disease

L Zell,^{*MD}, P Scheffler,^{**MD}, M Heger,^{***MD}, A Steuckardt-Götz,^{+MD}, A Buchter,^{++MD}

Abstract

Introduction: Primary thrombosis of the upper extremity (Paget-Schroetter syndrome, effort thrombosis) is usually not accepted either as a work-related accident or an occupational disease by the industrial injuries insurance authorities. This study aims to look at the circumstances under which this kind of thrombosis may be recognised as an occupational hazard. **Materials and Methods:** After thoroughly studying the clinical records of 82 patients with thrombosis of the upper extremity in the outpatient department for angiology during a 10-year period, we found that in 51 cases the thrombosis was caused by secondary reasons (i.e., central venous catheterisation, malignoma, hypercoagulability, postoperative). In the remaining 31 patients, we painstakingly conducted a general and occupational history and further clinical, laboratory or technical investigations to differentiate the aetiology. **Results:** Twenty-eight of 31 patients with suspected primary thrombosis of the axillary or subclavian vein (Paget-Schroetter syndrome) showed unusual private or occupational physical exercise as the underlying cause. Sixteen patients reported only short physical exercise up to several hours or acute trauma. Longer physical exercise periods (several days, months or even more) were found in 12 patients. Ten of the 28 patients who acquired the thrombosis doing occupational tasks covered by the industrial insurance, may be eligible for compensation. In 4 of these 10 insured cases, we found strong hints for work accidents, 6 patients of this group apparently revealed criteria of occupational diseases. Four of the 28 patients had a combination of physical effort and additional thrombogenic risk factors (cardiac pacemaker, hypercoagulability). **Conclusion:** Primary thrombosis of the upper extremity (Paget-Schroetter syndrome) should be announced to and compensated by the industrial injuries insurance either as work-related accident or occupational disease if the patient's occupational history shows close relationship between thrombosis manifestation and extraordinary physical efforts or exercises during occupational strains. For this reason, we recommend the recognition of the Paget-Schroetter syndrome in the national list of occupational diseases.

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Introduction

Concerning patients with thrombosis of the upper extremity, you can find only a small number of case studies regarding the question of assessment in occupational medicine.¹⁻³ This study aims to look at the circumstances under which this kind of thrombosis may be recognised as an occupational hazard.

A synonym for such kind of thrombosis of the upper extremity is the Paget-Schroetter syndrome. This name originates from first descriptions of this disease by the

well-known English physician, Sir James Paget, after whom several other diseases and syndromes have been named.^{4,5} The other was Leopold von Schroetter from Austria, who described a patient with thrombosis of the subclavian vein in 1899.^{5,6} In 1949, the English surgeon, Hughes, searched literature for cases of thrombosis of the upper extremity and gathered 320 cases in a detailed case study, further describing a comprehensive range of triggering physical strain effects. He also made the proposal to replace the until then common medical terms of "primary"

* Head of Department

EnBW Energie Baden-Württemberg AG, Arbeitsmedizinischer Dienst, Karlsruhe, Germany
Institut und Poliklinik für Arbeitsmedizin der Universität des Saarlandes, Homburg, Germany

** Professor and Head of Department

Medizinische Klinik, Innere Medizin III, Abteilung für Angiologie, Homburg, Germany

*** Head of Department

Landesamt für Arbeitssicherheit, Immisionsschutz und Gesundheit, Abteilung Medizinischer Arbeitsschutz und Gesundheit, Saarbrücken, Germany

+ Resident

++ Professor and Director

Institut und Poliklinik für Arbeitsmedizin der Universität des Saarlandes, Homburg, Germany

Address for Reprints: Dr Lothar Zell, EnBW Energie Baden-Württemberg AG, Arbeitsmedizinischer Dienst, Durlacher Allee 93, D-76131 Karlsruhe, Germany.

E-mail: l.zell@enbw.com

and “spontaneous” thromboses in the axillary and subclavian veins by the eponym “Paget-Schroetter syndrome”.^{5,7}

A lot of other very common and well-known synonyms have been used up to today. For example, the terms “primary” thrombosis, “spontaneous” thrombosis or “idiopathic” thrombosis. All these synonyms indicate that the cause for this kind of thrombosis was unknown, or could not be found, or perhaps the history of the patients was not sufficiently researched. Another common synonym used is the term “effort thrombosis”. This very common term indicates a causal relationship between an extraordinary physical strain and the aetiology of this thrombosis.⁸⁻¹² Still it is surprising that this syndrome was never discussed in occupational medicine and literature.

Pathophysiologically, this thrombosis is usually caused by a single or repeat stretching of the subclavian or axillary vein; for example, during extreme motion or strenuous activities. Such stretching of the vein can lead to traumatic fissures in the venous intima which is the inner layer of the venous vessel. These microtraumatisations of the venous intima are associated with local activation of coagulation and adhesion of thrombocytes. By this way, an appositional increase of a thrombus up to a total or subtotal occlusion of the venous vessel is possible. Additionally, recurrent microtraumatisations caused by repeated extraordinary physical strain can especially lead to the development of a thrombosis. In addition, anatomical compression of the subclavian vein by surrounding bones, ligaments or muscles structures with a venous stasis and a resulting activation of coagulation may play an important role in the pathophysiology of thrombogenesis.^{11,13}

Clinical symptom of this disease includes swelling of the upper extremity which develops immediately after causal physical strain or after a delay of several hours or days. The same is true for symptoms like pain, cyanosis or paraesthesia. Mostly, it is only after a few days or weeks that prominent superficial collateral veins develop (Figs. 1 and 2).

Correct diagnosis, history and clinical findings of the reported symptoms are important. Duplex scans are the preferred method for final diagnosis and for the last few years, these scans have substituted phlebography. Phlebography is used only in cases of unclarified findings (Fig. 3).

The following clinical case demonstrates the actual question and the reasons that lead us to research aspects of Paget-Schroetter syndrome in occupational medicine as an occupational disease or a work-related accident.

A 34-year-old male patient was transferred to our emergency unit with symptoms of thrombosis of the subclavian vein. He had never done any heavy physical work before and had been unemployed for the last 4 years.

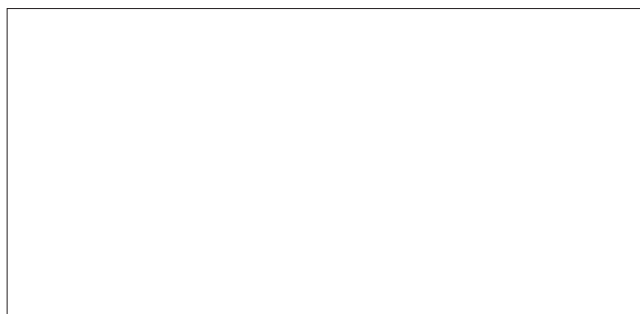


Fig. 1. A very rare case of Paget-Schroetter syndrome on both sides. Infrared photography showing superficial collateral veins.

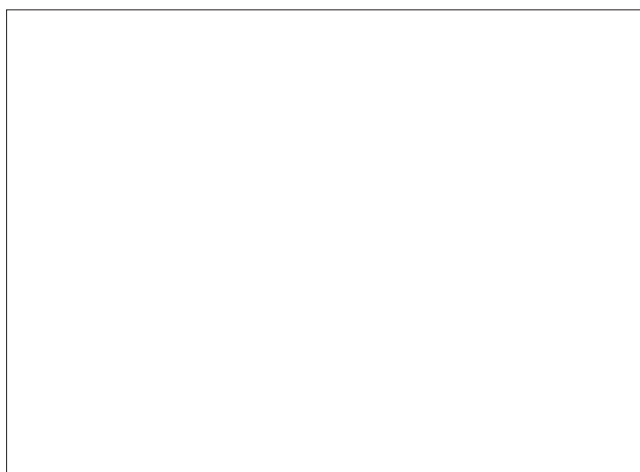


Fig. 2. Paget-Schroetter syndrome: scar after thrombectomy.

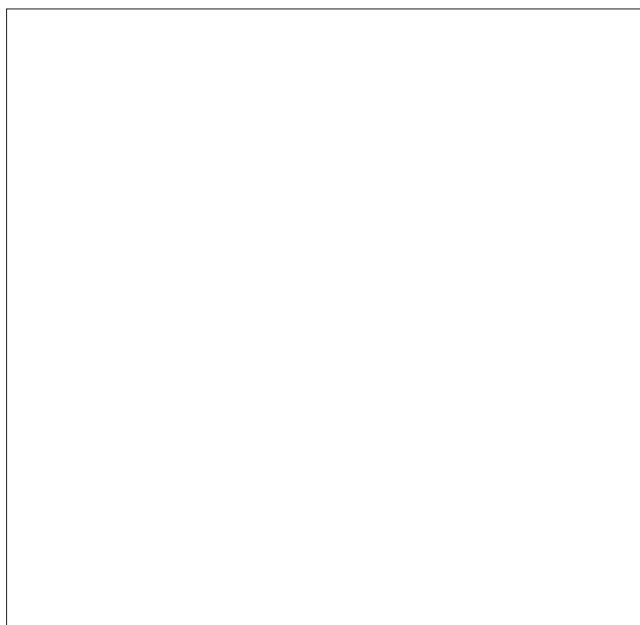


Fig. 3. Phlebography: thrombosis in the subclavian vein on the right-hand side with disruption of contrast medium.

He had just started a new employment where he had to bark logs the whole day with a barking knife. After working on the job for 4 days, the first symptom of thrombosis of the

upper extremity appeared. We referred this patient to the industrial injuries insurance for compensation and reported it as an occupational disease or a work-related accident. The insurance company refused to recognise this disease with the reasoning that this disease had not been described and discussed in the occupational health literature, and that there was no experience in occupational medicine concerning Paget-Schroetter syndrome. This motivated us to further research this disease in a larger group of patients.

Material and Methods

The records of 82 patients suffering from thrombosis of the upper extremity were reviewed. Records of these patients had been gathered for a period of 10 years at the outpatient department for angiology in our hospital. In 51 patients, the thrombosis could clearly or exclusively be classified as a consequence of “secondary” causes (central venous catheter, malignant neoplasms, hypercoagulability, effects following surgery, cervical rib^{14,15}). In the other 31 patients, whose records showed extraordinary physical strain, an unspecified cause or no cause at all for the genesis of a thrombosis, were examined either at home or at the outpatient department of our hospital and institute (Figs. 4 and 5). On this occasion, case histories were scrutinised in retrospect, prior medical diagnostic findings were gathered and in some cases additional examinations were carried out (particularly in order to complete and verify possible “secondary” causes).

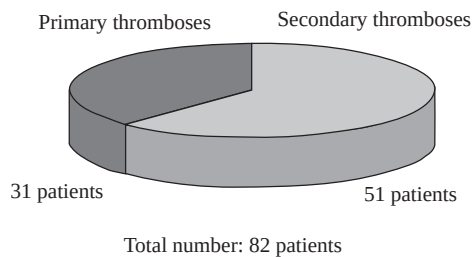


Fig. 4. Investigating the history of 82 patients with thrombosis of the upper extremity over a 10-year period.

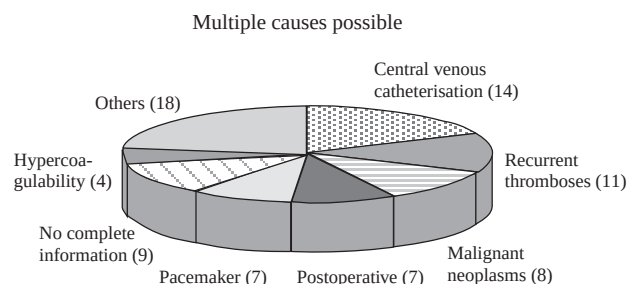


Fig. 5. Variety of causes in the group of patients ($n = 51$) with secondary thrombosis of the upper extremity.

Results

Investigations of 31 patients who had indications of a primary or effort thrombosis showed the following results:

Concerning the question of an extraordinary physical strain in relation with the beginning of first symptoms, 16 patients had an acute trauma or a strained exercise up to 1 day. Twelve patients showed unusual physical strain for several days or weeks (Fig. 6). The differentiation of the duration of up to 1 day or for several days is important in the German industrial injuries insurance; as in Germany, a work-related accident is usually defined by a duration of causal damage up to 1 day while an occupational disease is defined by a longer duration of causal damage.

All the 28 patients with extraordinary physical strain were examined for other additional causes or risk factors for a thrombosis. Four patients showed supplementary risk factors apart from the physical strain. In 1 case, there was cervical rib; in 2 cases, a hypercoagulability; and in another case, a cardiac pacemaker (Fig. 7).

Additionally, it would be interesting to see how many of these 28 patients performed this extraordinary physical strain in the course of their leisure or private activities or during employment covered by industrial injuries insurance. The thrombosis of approximately one third or 10 patients was caused by employment covered by insurance (Fig. 8).

Most of the patients had a lot of residual complaints and disabilities even after years of the first manifestation of

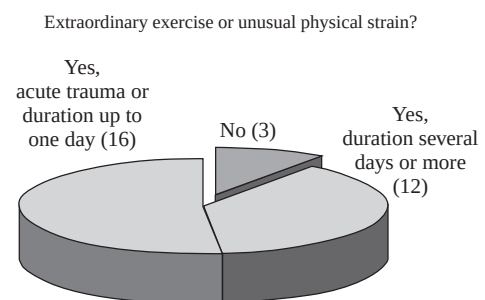


Fig. 6. Thirty-one primary thromboses of the subclavian/axillary vein.

Thrombosis caused by unusual physical strain and additional pathogenetic risk factors?

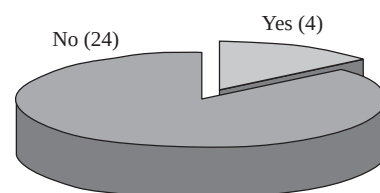


Fig. 7. Twenty-eight patients with thrombosis caused by extraordinary physical strain.

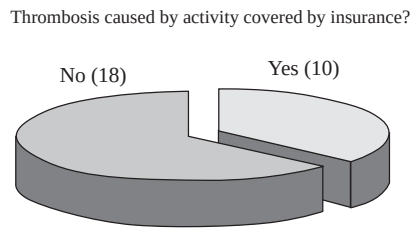


Fig. 8. Twenty-eight patients with thrombosis caused by extraordinary physical strain.

their thrombosis, especially problems or symptoms such as swelling and pain during working or when doing physical activities. Another residual symptom was protruding superficial collateral veins. One patient had a complicating pulmonary embolism.

Some examples for the causative physical strain of some patients are: In the group with short-term strain, we have an example of the jerky lifting of a heavy oven, or the catching of a falling patient in a hospital by a radiographer. We had 2 patients with an acute trauma caused by wrestling.¹⁶ Other examples include unusual sport exercises.¹⁷ Three of these patients have already been recognised by their industrial injuries insurance as a work-related accident.¹⁸

Long-term physical strain results in recurrent micro-traumatisation of the venous intima which also leads to thrombosis. Among the activities are—overhead work, e.g., the scraping of wallpaper off the ceiling for several days during renovation work; installing overhead panels; adjustment of vehicle tracks overhead during excessive workload or the suspension of heavy electric cables overhead in underground coal mining.¹⁵

Conclusion

In Germany and many other countries, Paget-Schroetter syndrome has not been discussed in occupational health. There are no experiences regarding the assessment of the Paget-Schroetter syndrome as an occupational disease or a work-related accident but pathogenetically, there exists an ascertained relationship between physical strain and the disease caused by lesions of the venous intima. In conclusion, we recommend that physicians report cases with this kind of thrombosis to the industrial injuries insurance if the causative activity is covered by insurance. It is obvious that further research and discussion of the Paget-Schroetter syndrome in occupational health and accident insurance are necessary. For this reason, we

recommend the recognition of the Paget-Schroetter syndrome in the national list of occupational diseases.

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