however, requires modification in view of the desperate condition of the patient. It is useless to attempt suture of the perforation, as the stitches will fail to hold in the diseased bowel wall, and, in any case, further perforations are usually impending at other sites in the colon. The caecum and colon as far down as the lower third of the pelvic colon must be excised, the distal end of the latter being brought out as a colostomy. Anastomosis of the ileum to the pelvi-rectal region would add a further quarter of an hour to the operation, and as the patient is unlikely to withstand this prolongation of operative time the severed end of the ileum is brought out as an ileostomy. Restoration of continuity of the bowel is delayed until the patient has fully recovered from the initial operation.

Discussion

I cannot agree with Counsell and Goligher (1952) in accepting the possibility of malignant change in ulcerative colitis as the dominating consideration in its surgical management. When true adenomata or stricture formation are present, then there is no question of the nature of the surgery that must be undertaken, but to advocate the removal of the rectum to prevent the formation of cancer when there is no certainty of its development is, I consider, too radical advice. The five specimens concerned in this article have been searched for cancer change and none has been found. It is unlikely to exist in the residua of the large bowels which have been retained, in view of the great improvement in the patients' general condition and the subsidence of the inflammatory changes, confirmed by proctoscopy. That the disease may recur in the rectum is a possibility, but the short follow-up of the present series has shown, so far, no indication of such a relapse. Even should a proportion have a return of symptoms, the establishment of an ileostomy and the excision of the rectum and terminal colon is no great surgical feat, and it can surely be reserved for those for whom it is a necessity rather than condemn all and sundry to a permanent ileostomy.

I have remarked on the noticeably smooth post-operative recovery of these patients, and from my own experience with ileostomies I have no doubt whatsoever about which is the easier. On this ground alone the operative mortality is likely to be very much less than the considerable mortality that accompanies ileostomy, and is an added argument in its favour.

Frequency of bowel action is unavoidable, but it is a small price to pay to escape an ileostomy life. In the second patient reported in the series the caecum appeared only mildly affected with the disease, and it, instead of the small intestine, was anastomosed to the pelvi-rectal junction. This patient has only two actions a day, and that this modification can be applied in other cases with advantage is a possibility.

Summary

The preservation of the rectum and of the normal channel of evacuation is advocated in those cases of ulcerative colitis in which fibrosis, pelvi-rectal suppuration, or true adenoma formation is absent from this part of the large intestine.

A method whereby this can be carried out without a preliminary ileostomy is described. The ileum is divided and its proximal end is anastomosed to the pelvi-rectal region. The colon above the site of the anastomosis is cut across and both free ends are exteriorized. The distal colostomy opening is subsequently closed and colectomy performed.

Immediate subtotal colectomy is advocated as the operative method of choice in the fulminating case.

Cases are reported in which a return to good health has followed treatment by the methods described.

I would like to thank Dr. E. R. Cullinan, in whose charge these patients have been, for his collaboration and advice in this work; Dr. C. B. Lewis, who has been responsible for the anaesthesia in these cases; and the nursing and resident staff of the Gordon Hospital for their unfailing care and attention to the patients.

Since this paper was prepared the initial stages of the operation have been successfully carried out in four other patients. One of these now has her bowels open three times a day only.

REFERENCES

- Aylett, S. O. (1951). *Proc. roy. Soc. Med.*, **44**, 196.
 — (1952). *Ibid.*, **45**, 394.
 Corbett, R. S. (1952). *Ann. roy. Coll. Surg. Engl.*, **10**, 21.
 Counsell, P. B., and Dukes, C. E. (1952). *Brit. J. Surg.*, **39**, 485.
 — and Goligher, J. C. (1952). *Lancet*, **2**, 1045.
 Devine, H., and Devine, J. (1948). *British Medical Journal*, **2**, 127.
 Muir, E. G. (1947). *Proc. roy. Soc. Med.*, **40**, 831.

PHLEGMASIA CAERULEA DOLENS

BY

PETER MARTIN, V.R.D., M.Chir., F.R.C.S.Ed.

Surgeon and Lecturer in Surgery, Postgraduate Medical School of London; Surgeon, Chelmsford Hospital

Phlegmasia caerulea dolens occurs in the limbs as a result of what would appear to be complete or almost complete obstruction to the venous outflow. It is of sudden onset, and is often associated with extreme cyanosis, swelling, pain, and shock, resulting in circulatory stasis of such a degree that gangrene follows in approximately a half of all cases.

The condition is rare, but has attracted much attention, particularly in France, and has been called variously pseudo-embolic phlebitis, blue phlebitis (Grégoire, 1938), and gangrenous thrombophlebitis (de Vega and Andrés García, 1946). DeBakey and Ochsner (1949) suggested the name phlegmasia caerulea dolens, and this seems an adequately descriptive term. Since then further cases have been reported (Clain and Nussbaum, 1950; Haimovici, 1950; Veal *et al.*, 1951; Miles, 1951; Turner, 1952).

Causation

The condition may occur when the main vein of a limb is suddenly obstructed. Veal *et al.* described cases that resulted from prophylactic ligation of the inferior vena cava in ilio-femoral thrombophlebitis with or without embolism, and I have seen the most alarming cyanosis and swelling after ligation of the common iliac vein for repeated embolism.

On account of repeated pulmonary embolism a man aged 63 had the common iliac vein ligated, thrombosis being present in the femoral vein. The following day he was moderately shocked and complained of severe pain in the thigh and leg. The affected leg was grossly swollen, intensely cyanosed, and much colder than his fellow, the distal pulses being hardly felt. The leg was elevated to 45 degrees and reflex heating and tolazoline hydrochloride were given; within three hours the swelling had practically disappeared and a full bounding pulsation was felt in the dorsalis pedis and posterior tibial vessels.

More usually it would seem to follow thrombophlebitis, occurring spontaneously or after operation, or in the puerperium, or as a result of pressure or invasion of a vein by neoplasm, or in association with blood disease.

A woman aged 54 was admitted to hospital with a thrombophlebitis of the right leg. She had suffered from attacks of phlebitis in the right leg during the thirty months prior to admission, and two and a half years previously had had a splenectomy, at another hospital, for a supposed Banti's syndrome. She was treated in hospital with penicillin, heparin, and dicoumarol, but, as a result of haemarthrosis and epistaxis, anticoagulants had to be stopped. Five weeks later there was a sudden onset of severe pain, swelling, and cyanosis of the right leg.

On examination the patient was seen to be severely shocked, the right leg was intensely swollen and cyanotic up to the knee, and the distal half of the leg was cold and pulseless. The femoral pulse was palpable, but more distal pulses were not felt. A diagnosis of embolus was considered, but no origin was apparent, and it was thought that the ischaemia was the result of a massive venous obstruction.

Conservative treatment by elevation, reflex heating, and tolazoline hydrochloride was instituted, but after three days it was obvious that a patch of tissue in the sole of the foot was gangrenous. The swelling gradually subsided and the cyanosis diminished slightly. Investigation of the blood showed a polycythaemia vera, which was treated by radioactive phosphorus. After five weeks the gangrenous area, fortunately small and superficial in extent, sloughed and distal pulses became palpable. However, cyanosis and moderate swelling persisted and it became evident that all the superficial veins of the limb were thrombosed, and that there was a thrombosis of the inferior vena cava.

Six months later the patient was getting about, with the legs bandaged to control the swelling, and the pulses were all normal, a foot plethysmograph at this time showing a greatly diminished but adequate flow in the right foot with no response to reflex heating, thus indicating venous obstruction of such a degree that no reserve blood flow was available.

Massive venous thrombosis as a result of polycythaemia vera in this case caused such a degree of venous obstruction as to jeopardize the life of the limb. Swartley *et al.* (1942) reported a case occurring in an upper extremity, resulting in gangrene of the forearm.

Why only a minute proportion of cases of thrombophlebitis are followed by this train of alarming symptoms is not at present understood, and there are certainly other factors concerned apart from venous obstruction from thrombophlebitis.

Clinical Features

Clinically, the onset is one of dramatic suddenness and is associated with severe pain, sometimes of the greatest intensity. At the same time there is marked swelling of the limb, associated with shock which may even be fatal, and cyanosis occurs which varies greatly in degree, but which may be almost black. The arterial pulse may not be detectable in the distal parts, and the swelling of the limb will render its detection even more difficult. The limb rapidly cools off, and the appearances are similar to those of arterial embolism; this has led to exploration of the femoral artery on a number of occasions (Audier, 1935; Wertheimer and Friehe, 1935; Lindgren, 1937; and Sackenreiter, 1940). Gangrene may ensue, and may be of any degree from a superficial necrosis to a massive death of tissue demanding major amputation which is fatal in 25% of cases. On the other hand, the condition may settle down slowly, the swelling and cyanosis gradually disappearing, and the limb circulation returning almost to normal with or without a post-phlebotic syndrome of varying degree persisting. Sometimes recovery is as sudden as the onset, and it may be partial or complete. Death may occur at the onset from shock associated with loss of fluid into the affected limb and excessive pain, or it may occur later on as a result of gangrene. Pulmonary embolism is a common complication. There seem to be two main varieties, either swelling and cyanosis or arterial symptoms predominating, the latter simulating embolism. More often the types are mixed and there are signs of both venous and arterial occlusion.

It has been shown experimentally (Audier and Haimovici, 1938) that gangrene due to venous obstruction in an organ can be produced only if every vein draining that organ is ligated—in other words, if there is complete venous obstruction—and it may be that in some cases which proceed to massive gangrene this is in fact the mechanism. If this were so, recovery would seem impossible and recanalization could not occur in time to prevent massive death of tissue.

Illustrative Cases

As in fact most cases recover to a greater or less degree—sometimes with no loss of tissue, sometimes with minor gangrene—it seems to me that factors other than total venous obstruction must be significant in this syndrome.

A man aged 67 developed, on the fourth day after a partial gastrectomy, a sudden intense swelling of the left lower extremity up to the level of the groin, and the swollen limb became deeply cyanotic. He was severely shocked, and examination showed a palpable femoral pulse but no distal pulses, and the whole limb was much cooler than the unaffected limb. The next day, after a sudden pain in the chest, the patient died. Post-mortem examination revealed a pulmonary embolus as the immediate cause of death, and the affected limb showed a recent acute thrombophlebitis of the internal saphenous vein, the result of an infusion into this vessel, with no evidence of thrombosis of any other vein, and with no obstruction to the arterial tree.

In this case it seems that an acute superficial thrombophlebitis may have induced a spasm in either the veins or the arteries, or in both the veins and the arteries, of the limb, resulting in cessation of the circulation.

De Sousa Pereira (1946) found that where an isolated segment of vein is the site of thrombophlebitis there is accompanying venospasm extending far beyond the vein affected, and that this spasm can be relieved by anaesthesia of the vein wall or by interruption of the efferent pathways of the sympathetic chain. That veins can go into very effective spasm will be agreed by anyone who has had the experience of intravenous therapy or of operations on veins. Even varicose veins, where muscular walls are atrophied and often largely replaced by fibrous tissue, may be seen to contract.

Diminished oscillations of an artery in close proximity to a vein which is the site of thrombophlebitis has been demonstrated by DeBakey *et al.* (1939), and the association of arterial spasm with deep thrombophlebitis has been emphasized by Boyd (1947). Sutton (1952) reported a case of spasm of the brachial artery of such a degree as to render the hand and fingers bloodless after an intravenous infusion into the superficial median basilic vein at the elbow.

A woman aged 25, thirty weeks pregnant, was admitted to hospital complaining of a cramp-like pain in the left leg on movement. On lying in bed there was a dull ache in the limb together with a feeling of numbness. The limb was pulseless from the femoral artery downwards, there was a patchy cyanosis, and both leg and foot were cold. Sensation was normal and the toes could be moved voluntarily. The picture suggested an embolus, but there was no apparent origin for this, and the case was treated conservatively. Among other procedures, heparin was given via intravenous saline drip. On the third day, while I was examining the patient, she had a rigor, and the most remarkable change came over the affected limb, the cyanosis being replaced quite rapidly by an intense hyperaemia, the pulses throughout the limb becoming full and bounding, and the temperature of the limb gradually becoming higher than its contralateral fellow. Two weeks later the only abnormality was a mild post-phlebotic syndrome.

It seems that in this case an idiopathic deep thrombophlebitis initiated a severe degree of arterial spasm. No doubt sudden rigor was the result of impurities in the heparin and saline drip, and gave rise to a functional sympathectomy which itself "released" the spasm. Dissection of amputated limbs and post-mortem specimens has shown an apparently healthy arterial tree without any evidence of obstruction, though limbs removed by delayed amputation may show the distal arteries close to the gangrenous area filled with thrombus.

Discussion

In phlegmasia caerulea dolens the disappearance of the arterial pulse may be due to a reflex arterial spasm initiated by an irritative thrombophlebitis. On the other hand, when there is oedema and swelling of an extremity resulting from venous obstruction the digital pulse may become impalpable. In fact, in such a limb oscillations recorded by a sensitive galvanometer may be completely masked in spite of a normal blood flow. Clinically, therefore, disappearance of the distal pulses may not necessarily indicate any active spasm of the arteries, especially in those cases in which the swollen cyanotic type of affection predominates. There is no doubt that massive thrombosis of all, or the vast majority, of the veins of the limb results in gangrene, but in those cases in which recovery takes place partial venous obstruction by

thrombus is rendered temporarily complete by an associated spasm of the remaining veins in the limb. At the same time there may be some degree of arterial spasm as well, and it is these several factors, varying in degree, which give rise to the different clinical varieties.

The condition may well be of medico-legal importance, for the development of gangrene in a limb following operation for varicose veins does not necessarily mean that the main artery or even the main vein has been tied.

Much of the treatment is indicated by the pathology. Heparin should be given to achieve and maintain a clotting-time of 15 to 20 minutes, and associated shock is treated by transfusion and other methods. The affected limb should be elevated to an angle of at least 45 degrees to take the best advantage of gravity, and, if the patient's general condition allows, supervised active exercises should be vigorously pursued.

Veal *et al.* recommended this treatment in all cases, and considered that any sympathetic release should be avoided, as it increases the embarrassment of an obstructed venous system. They also stated that if the condition follows ligation of a main vein the ligature should be removed immediately.

If, however, the argument is accepted that the obstruction to the venous drainage is to some degree functional, then it would appear advisable to try to break this reflex by a method of sympathetic release, and, as I have indicated above, it seems to me that there must be a large measure of spasm, at least in those cases which have a chance of recovery.

In a heparinized patient any sympathectomy by resection or by paravertebral injection is dangerous and very likely to produce a retroperitoneal haematoma, and should therefore be avoided. A quite adequate sympathectomy can readily be achieved by means of heat applied to the trunk and unaffected limbs, liberal doses of alcohol, one of the best available vasodilators, and the induction of deep sleep with soluble amylobarbitone in doses of 3 gr. (0.2 g.) hourly until the patient is difficult to rouse, or liberal doses of morphine if pain is severe. This is a regimen appreciated by patients, and it has the advantage of allaying anxiety, besides achieving its effect on the sympathetic. The affected limb should be kept at room temperature, as should any limb threatened with gangrene, and intra-arterial tolazoline may be tried, although I have not been very greatly impressed with this procedure, and my own preference is for the intravenous route in urgent cases. As soon as the patient is fit enough, the elevated limb should be exercised actively several times a day under the supervision of a physiotherapist. In many cases the swelling resolves, the pain is relieved, and the circulation returns, often rather suddenly, in whole or in part. If return of circulation is incomplete, massive or superficial gangrene may result. Pulmonary embolism is a common danger, but heparinization will tend to prevent this complication. In the event of minor or major gangrene developing, removal of tissue (or amputation) should be restricted to the dead part, since the circulation in the surviving proximal segment has always completely recovered before the need for ablation of dead tissue arises.

Summary

The association of acute ischaemia or even gangrene with thrombophlebitis of the extremities is discussed, and the name phlegmasia caerulea dolens is considered to be adequately descriptive.

The different clinical manifestations are described.

The mechanism of the ischaemia is considered, and a theory advanced to account for the varieties.

Treatment is described in outline.

REFERENCES

- Audier, M. (1935). *Progr. méd.*, Paris, 1, 729.
— and Haimovici, H. (1938). *Presse méd.*, 46, 1403.
Boyd, A. M. (1947). *Manchr. med. Sch. Gaz.*, 26, 124.

- Clain, A., and Nussbaum, L. L. (1950). *Lancet*, 2, 290.
DeBakey, M., Burch, G. E., and Ochsner, A. (1939). *Proc. Soc. exp. Biol.*, N.Y., 41, 585.
— and Ochsner, A. (1949). *Surgery*, 26, 16.
de Sousa Pereira, A. (1946). *Ibid.*, 19, 731.
De Vega y Fernández-Crespo, R., and Andrés García, P. (1946). *Rev. clin. esp.*, 23, 306.
Grégoire, R. (1938). *Presse méd.*, 46, 1313.
Haimovici, H. (1950). *Circulation*, 1, 225.
Lindgren, S. (1937). *Upsala LäkFören. Förh.*, 42, 415.
Miles, R. M. (1951). *Surgery*, 30, 718.
Sackenreiter, G. (1940). *Presse méd.*, 48, 575.
Sutton, M. (1952). *British Medical Journal*, 2, 859.
Swartley, W. B., Weeder, S. D., and McLaughlin, E. F. (1942). *Ann. Surg.*, 116, 184.
Turner, D. P. B. (1952). *British Medical Journal*, 2, 1183.
Veal, J. R., Dugan, T. J., Jamison, W. L., and Bauersfeld, R. S. (1951). *Surgery*, 29, 355.
Wertheimer, P., and Frich, P. (1935). *Presse méd.*, 43, 1004.

SURVIVAL OF TRANSFUSED RED CELLS IN SCURVY

BY

C. MERSKEY, M.D., M.R.C.P.

Department of Medicine, University of Capetown

The pathogenesis of the anaemia of scurvy is obscure. One view is that the mechanism may be inadequate haemopoiesis due to deficiency of vitamin C. Vilter *et al.* (1946) and others have suggested that there may be a haemolytic process.

The theory of haemolysis has been based on the occurrence of reticulocytosis and abnormally high serum bilirubin and faecal and urinary urobilinogen values in patients with scurvy and anaemia. These findings are not necessarily due to haemolysis. A more reliable indication may be obtained by studying the fate of transfused red cells. As search of the literature has not revealed any report of such an investigation in scorbutic patients, studies of the survival of transfused cells have been made in eight patients with scurvy and anaemia and in one with scurvy but without anaemia.

In six of these patients the transfused red-cell survival time was greatly reduced, and this was interpreted as evidence of a haemolytic process.

Material and Methods

Eight adult male patients (seven Bantu and one Indian) who manifested severe scurvy and anaemia with normoblastic bone marrow have been investigated. The ninth patient, an adult male Bantu, was scorbutic but not anaemic. For some time before admission to the Groote Schuur Hospital these patients had been on a diet severely deficient in vitamin-C content, consisting almost solely of mealie meal (maize) and white bread. All presented the classical picture of scurvy, with spongy haemorrhagic gums, follicular hyperkeratoses, and intramuscular haematomata. There were no signs of any other disease.

The investigation of the eight anaemic cases was divided into four stages as follows:

(1) Initially the patient was kept at complete rest in bed, without any medication and on a diet entirely lacking vitamin C (mealie meal porridge without milk or sugar, stamped mealies, white bread without butter or jam, and black tea and coffee).

(2) After the short control period of rest in bed, the patient was given one transfusion of citrated bank blood which had been stored for at least 36 hours. Between 2 and 4 pints (1.1 and 2.3 litres) of blood were given in each case according to the degree of anaemia. The vitamin-C content of the blood used in each transfusion was not determined, but the average vitamin-C content of 8 pints (4.5 litres) of blood, taken and stored for 36 hours under the same conditions