An Objective Study of Lumbar Sympathectomy—II. Skin Ischaemia*

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Brit. med. J., 1966, 1, 943–947

Lumbar sympathectomy is often performed for ischaemia of the foot in patients in whom direct arterial surgery is not desirable.

In contrast to the muscle circulation, there is a high level of sympathetic vasoconstrictor tone of skin vessels in the foot, and their sympathetic denervation initially results in marked vasodilatation in both the experimental animal (Löfving and Mellander, 1956) and the normal human (Walker *et al.*, 1950).

However, in practice, the operation often fails to provide relief of peripheral nutritional insufficiency. Clinical studies indicate that rest pain is relieved in only 40 to 60% of cases (Berry *et al.*, 1955 ; Thimmig *et al.*, 1958 ; Gillespie, 1960a ; Taylor and Calo, 1962). The risk of development of gangrene appears to be reduced by sympathectomy (Edwards and Crane, 1961), but the efficacy for established gangrene is disputed: whereas Berry *et al.* (1955) claimed that sympathectomy allowed successful conservative amputation in 40% of patients, Taylor and Calo (1962) considered that it was rarely of benefit.

The factors resulting in ischaemic rest pain are not entirely clear. The pain appears to be of two types (Ross, 1953). One type is characterized by severe, deep, unremitting pain, and it may be that this results from ischaemic neuritis with demyelination of nerve fibres as described by Gairns *et al.* (1960). In the other type there is superficial burning pain which usually occurs when the foot is warmed in bed at night and is relieved by cooling and by dependency. Possibly, in this type heating causes an increase of skin metabolism greater than can be dealt with by the hyperaemic response to warming the foot, resulting in accumulation of metabolites causing pain. Alternatively, the development of rest pain with heating could result from diversion of flow, in the presence of a limited inflow, from nutritive pathways to arteriovenous channels, though this does not occur in normal subjects (Davis and Greene, 1959), or from superficial to deep pathways. Subsequent development of gangrene results from an inadequate inflammatory hyperaemic response to local trauma or infection. The severity of the trauma necessary to produce death of tissue will depend on the degree of ischaemia, but clearly the development of gangrene does not necessarily imply gross ischaemia of the whole foot.

From these considerations it would appear that the symptoms of skin ischaemia might often result from inability to develop a hyperaemic response to a variety of stimuli. Sympathectomy can be effective in relieving these symptoms only if part of the restriction of development of the hyperaemic response is due to the sympathetic vasoconstrictor tone. Sympathectomy will be ineffective if the impaired flow predominantly results from organic disease, either of collateral or of peripheral small vessels. The significance of small-vessel disease is disputed, though it is known to occur in association with diabetes (Megibow *et al.*, 1949 ; Mendlowitz *et al.*, 1953), with rheumatoid arthritis (Virtama, 1959 ; Soila *et al.*, 1959), and in hypertension (Mendlowitz, 1951 ; Folkow, 1956). Conrad and Green (1964) showed the peripheral resistance in the toes to be markedly increased in patients with ischaemic ulceration.

Even if an increased flow is achieved by sympathectomy, it will fail to relieve symptoms if they be due to irreversible changes—for example, ischaemic neuritis. Failure of relief will also occur if the increased flow is to arteriovenous communications rather than to the capillary circulation or if there is shunting of flow from distal to proximal areas.

The present study was undertaken to attempt to determine, by objective measurement of blood-flow in the foot and ankle, the frequency of failure of benefit from lumbar sympathectomy, the possible reasons for failure, and whether it is possible to predict the response to sympathectomy in individual patients.

Material

Thirty-five patients were studied before and after lumbar sympathectomy ; the operation was bilateral in 11 patients, so that 46 legs are considered. In all cases the legs were the site of occlusive peripheral arterial disease ; in 20 legs there were no symptoms of skin ischaemia, the operation having been performed for intermittent claudication ; rest pain was present in 21 legs, ischaemic ulceration in three, and digital gangrene in two.

All patients were studied by arteriography ; an occlusion was present in both the aorto-iliac and femoro-popliteal segments in nine legs, in the femoro-popliteal segment alone in 28, and in the distal vessels only in nine.

Direct arterial surgery was not initially thought to be justified in these patients because of an inadequate distal "run-off" or poor general condition of the patient.

Methods of Assessment

Clinical Assessment

Each patient was assessed before and six to eight weeks after surgery without knowledge of the results of the flow studies. Definition of genuine rest pain in the foot was occasionally difficult, and only patients with severe pain interfering with sleep were included in this category.

Failure of improvement of rest pain by sympathectomy was indicated by the need for subsequent amputation or attempted direct arterial surgery. The adequacy of sympathectomy was confirmed in all cases by histological examination of the chain and by a sweating test.

Plethysmography

The following measurements were made before and 10 to 14 days after sympathectomy:

1. Resting blood-flows at the ankle and forefoot under basal conditions with the leg exposed at a room temperature between 22 and 24° C. ; the ambient temperature was intentionally kept low to simulate normal environmental conditions when it might be expected that benefit from sympathectomy would be more apparent (Walker *et al.*, 1950).

2. Flows at the ankle and forefoot measured first during the reactive hyperaemia after a five-minute period of arterial occlusion, then following local heating of the leg by electric blankets to a temperature of 42° C. for 30 minutes, and finally during a combination of the two. For the reactive hyperaemia tests, measurements were made of the peak flow and time at which it occurred after release of the tourniquet. The peak flow during the combina-

* Part I ("Intermittent Claudication") appeared last week (p. 879).
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tion of local heating and reactive hyperaemia is referred to as the hyperaemia flow.

3. Perfusion pressure at the ankle by the method previously described (Myers and Irvine, 1966).

4. Peripheral resistance at the ankle from the ratio of the perfusion pressure to the hyperaemia flow at the ankle, as previously described (Myers and Irvine, 1966).

Technique

The strain-gauge plethysmograph was identical to that previously described (Myers and Irvine, 1966). Both gauges are considered predominantly to measure flow through skin and subcutaneous tissue, as flow through tendon, fascia, etc., is small (Cooper *et al.*, 1955). The non-circular cross-section of the forefoot does not invalidate the technique (Whitney, 1953). The venous occlusion cuff for measuring ankle-flows was placed at the knee and that for measuring foot flows at the ankle.

Symptomatic Response

Of the 21 legs subject to rest pain, 13 were relieved of pain ; the remaining eight gained no improvement, so that eventually amputation or an attempt at arterial reconstruction was necessary. In the five legs with ulceration or gangrene healing was achieved and major amputation avoided. Thus of the 26 legs with features of skin ischaemia 18 gained relief after sympathectomy.

Plethysmographic Findings

Relation to Symptoms (Table I)

Resting Flows.—The means of the resting flows before sympathectomy in both the foot and the ankle were approximately identical in the two groups. After sympathectomy there was a significant increase in the foot flows in each group ; the increases in ankle flows were considerably less marked. The post-sympathectomy flow was only slightly less the more severe the evidence of ischaemia. Of the 26 operations performed for symptoms of skin ischaemia, improvement of the foot flow by more than 0.5 ml./100 ml./min. occurred in 20, the post-operative value being up to 4.5 ml./100 ml./min. ; in the remainder flows were unchanged or slightly diminished, in no case by more than 0.5 ml./100 ml./min.

TABLE I.—*Flow Studies in Relation to Clinical Symptoms (Mean ± S.E.)*

Clinical Group		Foot Flows (ml./100 ml./min.) Resting	Foot Flows (ml./100 ml./min.) Hyperaemia	Ankle Flows (ml./100 ml./min.) Resting	Ankle Flows (ml./100 ml./min.) Hyperaemia	Peripheral Resistance at Ankle (Units)
No symptoms of skin ischaemia (20 legs)	Pre-	0·8 ± 0·1	3·4 ± 0·4	0·8 ± 0·1	3·2 ± 0·3	19·5 ± 2
	Post-	2·7 ± 0·3	4·6 ± 0·7	1·3 ± 0·1	3·7 ± 0·5	15·5 ± 1·5
Rest pain, ulceration or gangrene (26 legs)	Pre-	1·0 ± 0·1	2·1 ± 0·4	1·0 ± 0·1	3·4 ± 0·5	23 ± 2
	Post-	2·2 ± 0·2	3·5 ± 0·5	1·7 ± 0·2	3·1 ± 0·5	22·5 ± 2

Pre- = Pre-sympathectomy. Post- = Post-sympathectomy.

Hyperaemia Flows.—In contrast to the resting flows there was a marked difference between the groups in the pre-sympathectomy mean of the hyperaemia flows in both the foot and the ankle ; however, there was a wide overlap in the range of values in each group, and the values in individual patients did not reliably correspond to the clinical assessment of the nature and severity of symptoms. Following sympathectomy there was a significant increase in the foot flow in each group ; the increase in the ankle flow in the patients with no symptoms of skin ischaemia was considerably less, and in the patients with symptoms of ischaemia increase in the foot flow was accompanied by a slight decrease in the ankle flow. For the whole group of operations a decrease in the resting and hyperaemia flows at the ankle with an increase in the corresponding flows in the foot occurred in four cases, and similar changes occurred for the hyperaemia flows alone in a further six cases ; the reverse phenomenon of increase of ankle flows and decrease of foot flows after sympathectomy was not observed. There was little difference in the post-operative foot flows in the group with symptoms compared with the group without. Of the 26 operations for symptoms of skin ischaemia, increased hyperaemia flow of greater than 0.5 ml./100 ml./min. occurred in all but eight, flows being up to 9.4 ml./100 ml./min.

Peripheral Resistance.—The mean of the peripheral resistances at the ankle was higher in those patients with symptoms of skin ischaemia than in those without, the difference in the means being statistically significant ($t=2.72$, $P<0.01$).

Relation to Clinical Response (Table II)

Resting Flows.—The mean of the resting flow in the foot was lower in the "unchanged" compared with the "improved" group both before and after sympathectomy, and the difference in the post-sympathectomy values was statistically significant ($t=2.66$, $P<0.02$). In the clinically improved group, flows increased in all but three of the 18 operations, to up to 4.5 ml./100 ml./min. ; in the unchanged group an increase occurred in four of the eight legs to up to 2.9 ml./100 ml./min.

TABLE II.—*Flow Studies in Relation to Clinical Response (Mean ± S.E.)*

Clinical Response		Foot Flows (ml./100 ml./min.) Resting	Foot Flows (ml./100 ml./min.) Hyperaemia	Peripheral Resistance at Ankle (Units)
Improved (18 legs)	Pre-	1·2 ± 0·2	3·1 ± 0·4	20·5 ± 2·5
	Post-	2·6 ± 0·3	4·4 ± 0·6	22 ± 2·5
Unchanged (8 legs)	Pre-	0·6 ± 0·2	1·0 ± 0·2	29 ± 3·5
	Post-	1·3 ± 0·4	1·8 ± 0·5	24·5 ± 3·5

Pre- = Pre-sympathectomy. Post- = Post-sympathectomy.

Hyperaemia Flows.—The mean of the hyperaemia flows in the foot in the unchanged group was little higher than that of the resting flows, and the difference in the post-sympathectomy values in the unchanged and improved groups was again significant ($t=2.94$, $P<0.01$). All patients who failed to show an increase in the resting flow similarly failed to show an increase in the hyperaemia flow after sympathectomy.

Peripheral Resistance.—There was little appreciable difference in the means of the peripheral resistance at the ankle in the unchanged compared with the improved group.

Relation to Arteriography (Table III)

The pre-sympathectomy values of the means of the resting and hyperaemia flows showed little difference according to the site of the occlusion. However, the post-sympathectomy values were much higher in the group with distal disease than in the group with combined aorto-iliac and femoro-popliteal occlusions.

TABLE III.—*Flow Studies in Relation to Site of Occlusion (Mean ± S.E.)*

Site of Occlusion		Foot Flows (ml./100 ml./min.) Resting	Foot Flows (ml./100 ml./min.) Hyperaemia	Peripheral Resistance at Ankle (Units)
Aorto-iliac and femoro-popliteal (9 legs)	Pre-	0·8 ± 0·2	2·9 ± 0·7	26 ± 5
	Post-	1·6 ± 0·3	2·7 ± 0·8	20·5 ± 4
Femoro-popliteal only (28 legs)	Pre-	0·9 ± 0·1	2·8 ± 0·3	20·5 ± 1·5
	Post-	2·5 ± 0·3	4·1 ± 0·4	20·5 ± 1·5
Distal (9 legs)	Pre-	0·9 ± 0·2	3·4 ± 0·5	20 ± 3·5
	Post-	3·2 ± 0·4	5·5 ± 1·1	17·5 ± 4·5

Pre- = Pre-sympathectomy. Post- = Post-sympathectomy.

The mean of the peripheral resistances at the ankle was slightly higher in the group with aorto-iliac disease than in those with femoro-popliteal occlusions alone or with distal disease.

Prediction of Response to Sympathectomy

Efficacy of Pre-operative Vasodilating Technique.—In the patients with symptoms of skin ischaemia the mean of the hyperaemia flows in the foot before sympathectomy was 60% of that occurring after operation (Table I). Of course, the post-operative hyperaemia flow does not necessarily indicate maximal vasodilatation, though this is probably nearly so. Local heating or reactive hyperaemia alone before sympathectomy was not as effective as the combination, each producing only approximately 40% of the maximum post-operative hyperaemia flow in a group of 10 legs studied.

Post-operative Resting Flows.—At the time of testing, 10 to 14 days after sympathectomy, the resting flows were also only 60% as great as the hyperaemia flows, as a result of persistence or return of vasomotor tone (Table I).

Correlation of Hyperaemia Flows with Response (Fig. 1).—Comparison of the pre-operative hyperaemia flows with the post-operative resting flows in the foot showed a significant correlation ($t=3.04$, $P<0.01$). All but one of the patients who were considered on clinical grounds to have had a poor response to sympathectomy showed a pre-operative hyperaemia flow of less than 1.5 ml./100 ml./min.

Correlation of Time of Peak Flow with Response (Fig. 2).—There was also a highly significant correlation between the time of development of the peak flow in the foot after release of the arterial tourniquet, measured before sympathectomy, and the post-operative resting foot flow ($t=6.37$, $P<0.001$). All limbs which failed to gain clinical benefit had a delay in development of the peak flow of greater than 150 seconds.

Discussion

Relief of the symptoms of skin ischaemia by sympathectomy necessitates an increase of the blood-flow through the capillary circulation. Lynn and Barcroft (1950) demonstrated that the increased flow through the foot is maximal within 48 hours of the operation, but that flows fall sharply within the first seven to ten days. The improvement in flow is greatest at low environmental temperatures (Walker *et al.*, 1950; Tice *et al.*, 1963). General body-cooling results in increased sympathetic vasoconstrictor activity in the skin (Pickering, 1933), and this is abolished by sympathectomy (de Crinis *et al.*, 1959), though local cooling, down to 15° C., results in more pronounced vasoconstriction, which is only temporarily reduced by denervation (Freeman, 1935).

The findings in this study that the resting flows in the foot, measured at 10 to 14 days after surgery with the limb exposed in an environmental temperature of 22 to 24° C., are increased even in the presence of severe symptoms of ischaemia, confirm the studies of McPherson and Kessel (1956) and Gillespie (1960b).

However, it is considered that the symptoms of rest pain and the failure of healing of ulcerated or gangrenous tissue result from inability to develop an adequate hyperaemic response in the area concerned. The demonstration in these patients that the flows in the foot during a combination of local heating and reactive hyperaemia are also increased after sympathectomy, again even in patients with severe ischaemia, lends further support to the rationale of the operation. The importance of impairment of the hyperaemic response in the production of symptoms is supported by the finding in this study that the mean resting flows in the foot before operation were equal in the groups with and without symptoms of skin ischaemia, but that the hyperaemia flows were much lower in the latter. This, together with the difference in the peripheral resistances in the ankle in the two groups, suggests that the impairment of the hyperaemic response may result from either proximal disease or distal small-vessel organic disease. The hyperaemia flows at the ankle were not increased in the group with symptoms of skin ischaemia. In 10 of the 46 legs in the whole group there was a decrease in ankle flow with an increase of foot flow, suggesting a shunting of flow from the skin of the leg to the foot (Hyman and Winsor, 1959). The three patients with ischaemic leg ulceration appeared to gain benefit from the operation, but this cannot always be expected with a standard sympathectomy.

The relatively poor response of the patients with combined aorto-iliac and femoro-popliteal occlusions has also been described by Hill *et al.* (1962). It probably results from the presence of more extensive disease in these patients, both proximally and distally.

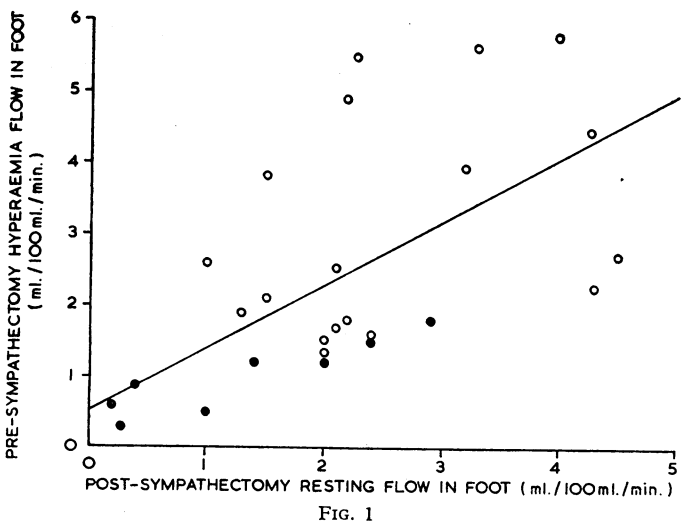

FIG. 1

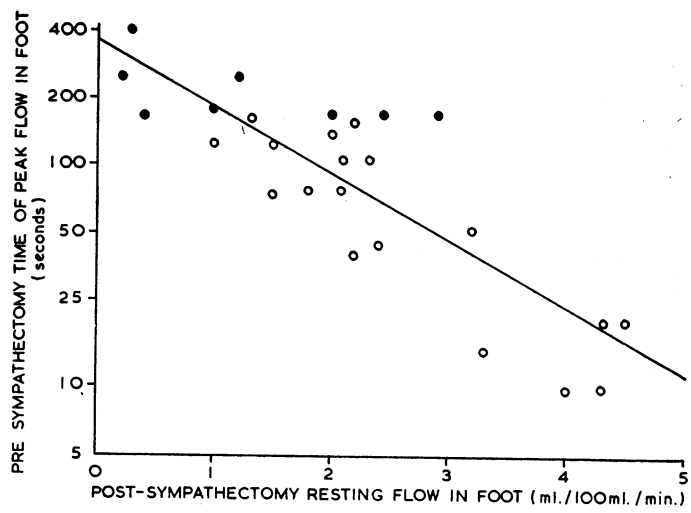

FIG. 2

FIG. 1.—Graph illustrating prediction of response to sympathectomy from pre-sympathectomy hyperaemia flow in foot; there is a significant correlation between pre-sympathectomy hyperaemia flows and post-sympathectomy resting flows ($r=0.58$, $t=3.04$, $P<0.01$); all but one of the patients who failed to gain clinical improvement from operation showed hyperaemia flow of less than 1.5 ml./100 ml./min. (○=clinically improved; ●=clinically unchanged.) FIG. 2.—Graph illustrating prediction of response to sympathectomy from pre-sympathectomy time of peak flow during hyperaemia in foot; there is a significant correlation between the logarithm of the pre-sympathectomy times of peak flow and the post-sympathectomy resting flows. ($r=-0.80$, $t=6.37$, $P<0.001$); all patients who failed to gain clinical improvement from operation showed delay in development of the peak flow of more than 150 seconds. (○=clinically improved; ●=clinically unchanged.)

Eight of the 26 limbs with symptoms of skin ischaemia failed to benefit from sympathectomy. There are several possible reasons for this. Probably the most important factor limiting the response is the presence of severe restriction of arterial inflow by poor development of collateral vessels, which are probably not greatly influenced by sympathectomy (Myers and Irvine, 1966). An alternative factor is the presence of distal small-vessel disease which reduces the degree of vasodilatation following denervation. In the limbs that were not improved the mean hyperaemia flow was much lower than in those that gained benefit, being little greater than the resting flow, whereas there was little difference in the peripheral resistance at the ankle, suggesting that limitation of proximal inflow was the more significant factor.

The presence of irreversible changes in peripheral tissues, for example associated with ischaemic neuritis, may have been the reason for failure in the four legs in which there was persistence of severe rest pain in spite of a considerable increase in resting flows. Alternatively, the increased flow in these patients might have resulted from the opening of arteriovenous pathways, shunting flow away from nutritive channels (Freeman *et al.*, 1947): sympathectomy causes a marked increase in venous oxygen saturation (de Takats, 1958), but in the normal subject sympathetic block results in a decrease of the oxygen tension in the skin (Davis and Greene, 1959), suggesting preferential opening of arteriovenous pathways resulting in stagnation of capillary flow. It is not known whether this decrease in skin oxygen tension also occurs in the presence of arterial disease. Shunting of flow from distal to proximal areas of the leg is a theoretical possibility, but no patient in the group showed a significant fall in resting flow in the foot after operation; in fact, the reverse phenomenon, of shunting of flow from the skin of the leg to the foot, often occurred. Finally, deterioration might result from further vascular occlusion either by an embolus dislodged from the aorta or by peripheral thrombosis induced by an episode of operative hypotension, or by post-operative disturbances in the clotting mechanisms, and this occurred in two patients who were studied but were not included in this report.

Occasionally assessment of whether there are genuine symptoms of skin ischaemia sufficient to warrant surgery is difficult, and a reliable objective index of the need for sympathectomy would be most helpful in these patients. This would at least require demonstration of a clear difference in the range of flows between patients with and patients without symptoms of skin ischaemia, but it has not been possible in this study to demonstrate such a distinction. The decision whether the incapacity warrants consideration of sympathectomy is perhaps best made on clinical grounds.

Once this decision is made it is generally agreed that there are no reliable clinical criteria to indicate which patients will benefit from the operation. A vasodilating technique was used to attempt to simulate the effect of sympathectomy so as to enable pre-operative prediction of the response to be made. In particular, it was desired to develop a method by which to predict reliably which patients would not respond, so allowing avoidance of an unnecessary operation. Several different techniques for this purpose have been described: measuring skin temperatures or blood-flows after reflex heating (Husni and Simeone, 1957; Smithwick, 1957), peripheral nerve-block (Husni and Simeone, 1957), sympathetic block (Thimmig *et al.*, 1958), epidural, spinal, or general anaesthesia (Holopainen, 1963), or vasodilator drugs (Knox, 1959). The disadvantage of many of these tests is that failure to release sympathetic tone distally, or reduction of the systemic blood-pressure, will result in prediction of a poor response that will not be fulfilled, and they are generally considered to be unreliable (Freeman *et al.*, 1947; Husni and Simeone, 1957; Edwards and Crane, 1961).

The effect of local heating and reactive hyperaemia is much more predictable, and "false negatives" are less likely. It has been shown that, alone, either produces a greater hyperaemia flow than sympathetic block, reflex heating, or intra-arterial vasodilator drugs (Wright and Phelps, 1940; Stein, 1956), and the combination of the two in this study produced an even greater response. However, the effects of heating and of denervation on the skin micro-circulation in the normal subject are not identical. Whereas denervation results in a decreased tissue-oxygen tension, associated with an increased venous-oxygen saturation suggesting preferential flow through arteriovenous channels, heating causes an increased skin-oxygen tension indicating increased capillary flow (Davis and Greene, 1959). Moreover, reactive hyperaemia actually induces selective constriction of arteriovenous shunts with secondary capillary dilatation (Burch and De Pasquale, 1962). Thus this test provides an index of the ability to increase capillary flow. The development of a good hyperaemic response before operation does not necessarily imply that the patient will obtain a good response to sympathectomy, but in this study a poor response to surgery was always associated with a virtually absent pre-operative hyperaemic response.

Not only the peak flows but also the delay in their development have been held to be of significance (Smithwick, 1957), and this was also confirmed in our study; but the mechanism of this delay is not clear (Myers, 1964), and its value for prediction of the response is uncertain.

Summary

Thirty-five patients having 46 operations were studied before and 10 to 14 days after lumbar sympathectomy by strain-gauge plethysmography. The operation usually resulted in a significant increase in blood-flow in the foot, both when cooled and following a combination of local heating and reactive hyperaemia. The response was less the more severe the symptoms and the more proximal the site of disease. Rest pain probably results from an inadequate hyperaemic response to warming of the foot, and the finding of an increased hyperaemia flow after sympathectomy adds support to the rationale of the operation. Restriction of the hyperaemic response may result from either proximal or distal organic disease.

Eight of 26 legs with symptoms of skin ischaemia failed to benefit from the operation. Probably the most important cause was severe limitation of inflow through collateral vessels rather than distal disease. In some patients failure may have been due to the presence of irreversible distal changes, to the occurrence of shunting to arteriovenous channels, or to precipitation of complete occlusion of a previously stenosed vessel. There was no evidence of shunting of flow from distal to proximal tissues, though the opposite phenomenon commonly occurred.

A technique of local heating and reactive hyperaemia was used to assess the degree of hyperaemia which could be achieved in the foot, for the purpose of predicting the response to sympathectomy. There was a high degree of correlation between the peak flow during hyperaemia and the time of its occurrence before sympathectomy with the post-operative resting flow and with the clinical response; all but one of the patients who failed to gain relief of symptoms of skin ischaemia following the operation had shown a pre-operative hyperaemia flow of less than 1.5 ml./100 ml./min. and a delay in development of the peak flow after release of the arterial tourniquet of more than 150 seconds.

We wish to thank Professor M. R. Ewing for his support of this study. We also wish to thank Mr. T. Needham and Mr. M. England for technical assistance. One of us (K. A. M.) was supported by a Smith and Nephew Fellowship.

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Clinical Features of Autosomal Dominant and Sex-linked Ichthyosis in an English Population*

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Brit. med. J., 1966, **1**, 947–950

It is generally believed that ichthyosis is a relatively common hereditary disorder, though there is little information on its actual prevalence. Rare varieties have been adequately defined in descriptive terms, but difficulties have arisen over the characteristics of frequently encountered forms. In order to obtain information on prevalence, clinical features, and genetics of ichthyosis, a survey was undertaken in the five counties surrounding Oxford. Through studying clinical features of ichthyosis in families where the mode of inheritance could be defined it was possible to separate two distinct varieties due to either autosomal dominant or to sex-linked genes. As these two conditions account for over 95% of all cases of ichthyosis, the characteristic clinical features of each variety are emphasized. A genetical classification of ichthyosis is given in Table I.

TABLE I.—*Genetical Classification of Ichthyosis*

Genetical Group	Varieties	Qualifying Terms used in Descriptive Classification
Autosomal dominant	(a) Autosomal dominant ichthyosis	Ichthyosis nacrée, nitida, vulgaris, or simplex; xerodermia; pityriasis vulgaris
	(b) Bullous ichthyosiform erythrodermia	Hyperkeratose ichthyosiforme
	(c) Ichthyosis hystrix (Lambert family)	Ichthyosis cornea
Autosomal recessive	(a) Ichthyosiform erythrodermia	Harlequin foetus. Ichthyosis congenita, larvata, tarda, mitis, or inversa
	(b) Lamellar ichthyosis	Collodion baby
	(c) Refsum's syndrome	
	(d) Sjögren-Larsson syndrome	
Sex-linked recessive	Sex-linked ichthyosis	Ichthyosis serpentina, saurodermia, nigricans, or vulgaris

Historical Background

Although known to Avicenna as "albarras nigra" and also described in the fourteenth century by the Chinese (Chan, 1950), ichthyosis was first clearly described in the English literature by Willan in 1808. Many different varieties were subsequently reported, but most authors recognized two major categories. One was characterized by small, fine scales with "a greyish, silvery and glossy" appearance (Erasmus Wilson, 1857), previously likened by Alibert (1806) to mother-of-pearl (ichthyosis nacrée). In the other category the scales were larger, "dark-coloured, greyish green, brown, or black" and more firmly fixed to the underlying skin (Hebra and Kaposi, 1874). Radcliffe Crocker (1896) gave a detailed account of the clinical features and in contrast to most previous reviewers noted that "fanciful names" merely led to confusion, and were to be avoided. Cockayne (1933) was the first to use a genetical classification, but neither he nor, more recently, Greither (1964) distinguished between the clinical characteristics of varieties with differing inheritance. Touraine (1958) was forced to construct many categories and subcategories to accommodate all recorded types of ichthyosis in a descriptive classification. Genetical and clinical evidence on which rare forms are classified as distinct entities in Table I is reviewed in detail elsewhere (Wells and Kerr, 1965).

Present Investigation

During 1962–4 persons with ichthyosis living in Oxfordshire, Northamptonshire, Buckinghamshire, Berkshire, and Wiltshire were ascertained from general practitioners, consultant dermatologists, hospital records, and school medical officers of health. An additional family with sex-linked ichthyosis was studied in Northern Ireland. Data for genetical and clinical analysis were sought from the families of 288 index cases and over 1,100 relatives were examined. Co-operation was excellent, as less than 1% of those approached refused to take part in the investigation.

* This paper represents part of a thesis by one of us (R. S. W.) accepted for the M.D. of the University of London.

† From the Population Genetics Research Unit, Oxford.

‡ Supported by the Postgraduate Medical Foundation, University of Sydney.