Summary of Cases of Adventitial Cystic Disease of the Popliteal Artery

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Adventitial cystic disease of the popliteal artery is explored. The results of correspondence with authors reporting this condition are elaborated upon. This has provided an opportunity to discuss the history of the condition, the findings in 115 cases which have come to the attention of the Correspondence Office dealing with this entity, and the results of treatment. A discussion of the suspected etiology of the condition is presented. The condition remains one of unknown etiology which can be treated by cyst evacuation or aspiration when the popliteal artery is patent and which is best treated by arterial reconstruction when the artery is occluded. The results of such treatment are good but are dependent upon technical excellence of the operative procedure.

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"UNUSUAL CASES ARE the spice of medicine." Barnett, Dugdale and Ferguson, 1966.[5]

Occlusion of the popliteal artery caused by adventitial cystic disease is rare. Therefore, during 1969 and again in 1975 and 1976, this office initiated correspondence with surgeons who had reported cases of this condition to obtain additional data which might cast light on the natural history of this condition, its etiology, relationship to systemic diseases elsewhere in the body, and results of treatment. The results of this correspondence were most gratifying. Newsletters were sent out in 1975 and 1976 summarizing the results of these surveys and providing up-to-date bibliographies which listed all known cases of this condition. Colleagues urged that this office prepare a summary for publication which would serve surgical science by providing a complete listing of cases and an accurate reference list. This report is such a summation. The fact that it is possible to prepare this presentation is due to scientific cooperation which allows surgeons on many continents speaking different languages to correspond with one another to provide information on a particularly interesting topic.

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Historical Introduction

Apparently, the first case of adventitial cystic disease of the popliteal artery was operated upon January 26, 1953 by Hierton[26] of the Department of Orthopaedic Surgery of the Norrbacka-Institutet, Stockholm, Sweden. The authors describe that a transverse incision was made in the middle of the thickened area of the artery and two thimbles-full of a mass resembling raspberry jelly emptied from an intramural, multilocular cavity. The condition was tentatively regarded as mucoid degeneration in the media and a saphenous vein graft was used to replace the affected segment of artery. Hierton discussed this peculiar vascular abnormality with Charles Rob, at that time Professor of Surgery at St. Mary's Hospital, London, and this resulted in a mutual publication in the *British Journal of Surgery* in 1957.[40]

In this paper, which described the four known cases encountered to that time, descriptive terms applied to the lesion included a "clear, jelly-like material similar in appearance to that seen in a ganglion," and "the specimen looked like a sausage and was 7 cm in length." "The lumen was compressed by an intramural cyst containing jelly under high tension." In this article, reference was made to the 1946 publication of Atkins and Key,[3] which had described adventitial cystic degeneration of the external iliac artery. In a subsequent publication, the lesion was described as reminiscent of a hotdog and a color photograph was provided.[39]

During the early 1960's, Ishikawa et al.[44] contributed an important diagnostic sign. They noted that normal distal pulsations were obliterated when the affected patient's knee was sharply flexed. This sign could be positive only in patients with arterial stenosis rather than total occlusion.

Jacquet and Meyer-Burgdorff[45] emphasized that, if

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the cyst had produced a highly stenotic lesion of the popliteal artery, a distal jet of blood flow might occur only at the very peak of systolic pressure. Eastcott[23] had noted this, suggesting that an arterial murmur over the popliteal fossa was an important sign in establishing proper diagnosis in young, nonsmoking patients who suffered from intermittent claudication.

In 1967, Taylor[90] made an important observation in relationship to his case, suggesting that "The sudden onset of symptoms in our case may have been due to the floor of the superficial cyst giving way, resulting in the extrusion of the contents into the dissection plane of the vessel, so forming an internal projection which constricted the lumen." Taylor, Taylor and Ramsay[90] thus felt that these cysts were similar to ganglia, which were degenerative cysts containing collagenous material and caused by trauma.

At the time of the first world survey of authors reporting this condition, it had been determined that 40 cases had been reported.[33] No systemic arterial or joint disease ever followed recognition of adventitial cystic disease of the popliteal artery. The incidence of this condition was approximately one in 1,200 cases of claudication,[52] or one in 1,000 femoral arteriograms.[56] Since then, correspondence has been maintained and at this time, there are 115 cases of cystic adventitial disease of the popliteal artery and an additional 21 cases of nonpopliteal cystic disease have also been recorded. The condition has been found in association with veins on three occasions (Fig. 1).

Profile of the Condition

Hierton and Lindberg[39] summarized the condition in a form which can hardly be improved upon although 20 years have passed since their initial observations. Their summary included: occurrence in young males, with sudden onset of cramps in the calf, development of typical intermittent claudication, presence of a localized stenosis and/or occlusion of the popliteal artery, absence of generalized arterial changes, formation of intramural cyst between media and adventitia compressing the arterial lumen, cyst contents of gelatinous material under tension, cyst wall lined by flattened cells, and structure of the cyst wall suggestive of mucinous degeneration.

To this can be added the lucid summation by Bliss in 1964,[9] a portion of which follows:

"*Etiology*: This is a condition of unknown pathogenesis which occurs principally in young adults

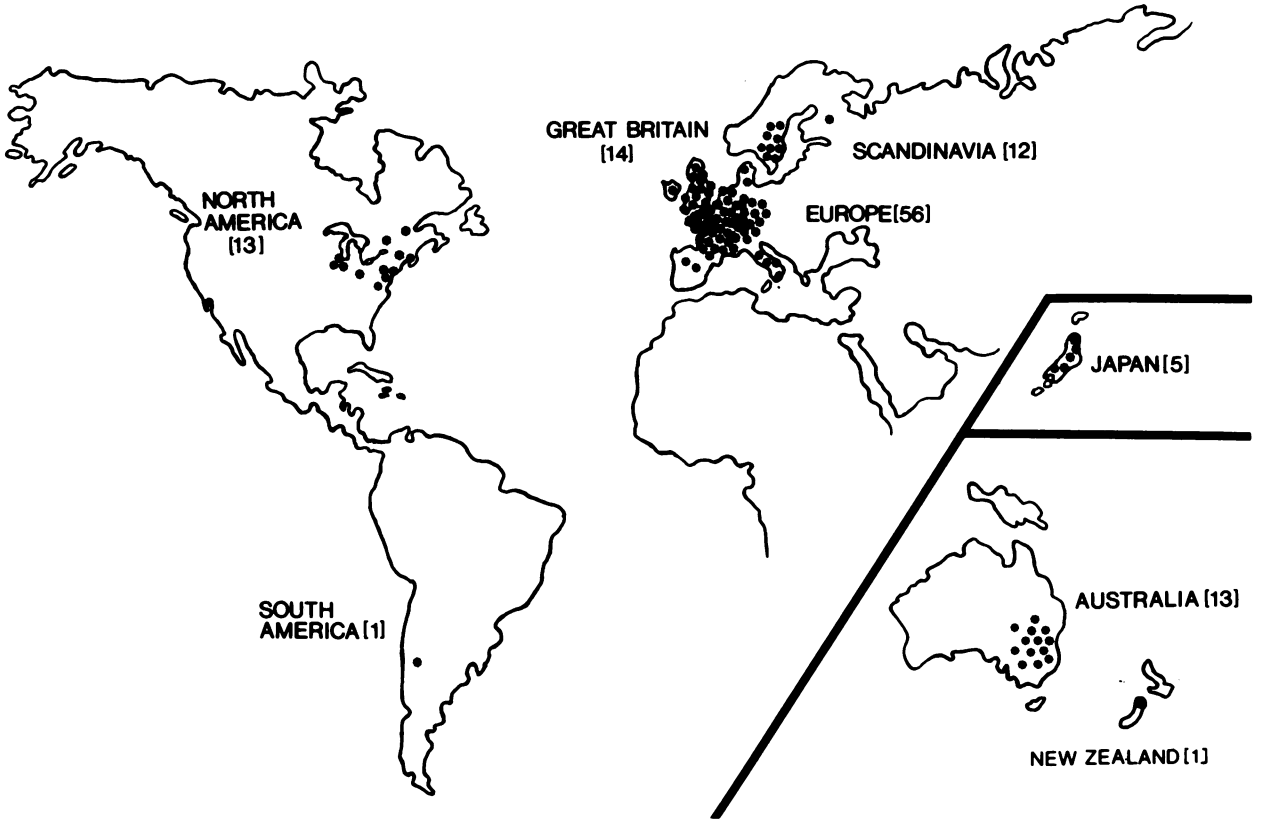


FIG. 1. World wide distribution of cases of adventitial cystic disease of the popliteal artery, indicating the paucity of such cases from the North American continent and the prevalence of such cases from Europe and Australasia.

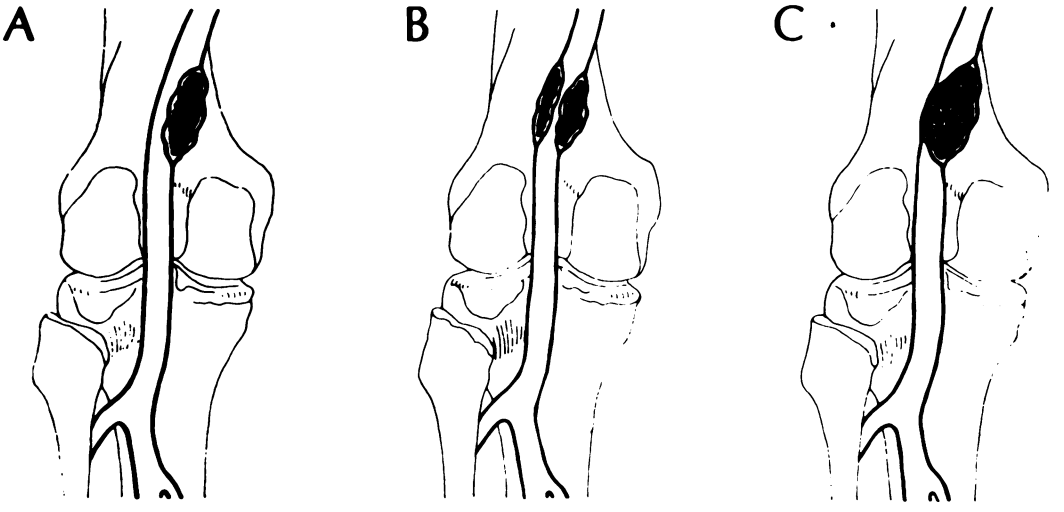


FIG. 2. These three diagrams indicate the configuration to be shown on arteriography in cystic adventitial disease of the popliteal artery. (A) shows the curvilinear scimitar sign when the cyst is medially or laterally placed in relation to the artery. (B) shows the hour glass configuration when the cyst encircles the arterial lumen. (C) shows total occlusion of the artery as the cyst enlarges to encroach upon the lumen itself.

employed in heavy manual leg work. Females and those in lighter occupations are occasionally affected. *Clinical picture*: There is a sudden onset of cramping pain in the calf, followed by the development of typical intermittent claudication. Signs of ischemia are present, although sometimes only on exertion, and may be exacerbated by full flexion of the knee. *Arteriography* shows a smooth-walled stenosis or complete block in the popliteal artery with an otherwise normal arterial tree (Fig. 2). *Pathology*: The unilocular or multilocular cyst is present within the wall of the popliteal artery compressing the lumen. *Treatment*: In most cases, evacuation of the cyst is sufficient. Thromboendarterectomy or incision and grafting may be necessary when secondary thrombosis of the lumen has occurred. *Prognosis*: The short-term prognosis after operation is excellent."

Summary of Known Cases

An extensive effort was made to obtain and translate into English all articles pertinent to the subject of cystic adventitial disease of the popliteal artery. This produced a total of 115 case reports (Table 1). Following this, an attempt was made to contact the senior author of each case report, asking him to provide information relative to the follow-up of his case and to comment upon the suspected etiology of the condition. A summary of these findings was circulated in a newsletter on two occasions, allowing authors to receive the viewpoints of other surgeons and providing a more complete bibliography. In this way, a more complete file, including cases previously unreported in the literature, was acquired for use in the Correspondence Office.

At the conclusion of this effort, it was possible to identify 105 cases which would be acceptable for inclusion in the following study. All cases which conformed to the description of cystic disease of the popliteal artery as described by Hierton[26] in his first publication were included. Those excluded were cases with inadequate documentation, those with involvement of arteries other than the popliteal and, of course, conditions such as hematoma of the popliteal space and other occlusions of the popliteal artery which were not pertinent to the study of this condition.

Particular attention was paid in review of the cases to age of the patient at time of presentation, sex, method of clinical presentation, methods of diagnosis used, methods and results of treatment employed, and theories of etiology. The follow-up information provided by the reporting author was accepted as factual with no further attempt made to contact the clinical patient.

In this group of 105 patients, there were 83 men and 18 women; in four instances of reporting, the sex of the patient was not stated. The mean age was 42 years, with a range of 11–70 years. Surgical procedures varied greatly but could be divided into cases treated by resectional therapy and those treated by a nonresection technique. There are many instances in which resection was done prior to knowledge of precise etiology of the arterial lesion. However, resection was performed principally if the popliteal artery was completely occluded or if there was extensive degeneration of the arterial wall. Nonresectional techniques were used when the occlusion was incomplete. In seven case reports, the type of surgical procedure was not identified with sufficient detail that it could be classified but, in the 98 remaining cases, 56 were treated by non-resectional techniques and 42 were treated by resection and replacement of the artery. Such replacement was accomplished by autogenous vein graft in 30 patients; synthetic graft replacement in seven; homograft replacement of arteries in two; and end-to-end

TABLE 1. *World Summary of Cases of Adventitial Cystic Disease of the Popliteal Artery*

No.	Author	Year	Age	Sex	Side	Arteriogram	Treatment
1	Ejrup & Hierton[26]	1953	32	M	L	Stenosis	Resection, vein graft
2	Hierton & Lindberg[39]	1957	25	M	R	Occlusion	Resection, homograft
3	Hierton & Lindberg[39]	1957	24	M	L	Occlusion	Resection, vein graft
4	Hierton & Lindberg[39]	1957	32	M	R	Occlusion	Resection, vein graft
5	Patel, Facquet & Piwnica[70]	1958	23	M	L	Stenosis	Evacuation
6	Tytgat, Derom & Galinsky[92]	1958	47	M	R	Stenosis	Resection, nylon graft
7	Andersson, et al.[2]	1959	48	M	L	Occlusion	Resection, homograft
8	Hierton*	1959	51	F	—	Stenosis†	Resection, vein graft
9	Holmes[42]	1960	42	M	L	Stenosis	Excision, no graft
10	Robb[75]	1960	39	M	R	Stenosis	Resection, vein graft
11	Delannoy & Martinot[19]	1960	38	M	R	Stenosis	Evacuation
12	Ishikawa, et al.[44]	1960	32	M	R	Stenosis	Resection
13	Chevrier[16]	1962	26	M	L	Stenosis	Evacuation
14	Marzoli, et al.[59]	1962	49	—	L	Stenosis	Excision, suture
15	Sutton[88]	1962	45	M	—	—	—
16	Bliss, et al.[10]	1963	40	F	L	Stenosis	Evacuation
17	Eastcott[23]	1963	48	M	—	Stenosis	Evacuation, endarterectomy
18	Simon[84]	1963	52	M	R	Stenosis	Resection, Dacron® graft
19	Lambley[50]	1963	47	M	L	Stenosis	Resection, Dacron graft
20	Patel & Cormier[69]	1963	50	F	R	Stenosis	Resection, repair
21	Gripe[32]	1963	37	F	—	Occlusion	Resection, vein graft
22	Vollmar[93]	1963	60	M	R	Stenosis	Resection, Dacron graft
23	Barnett & Morris[6]	1964	56	F	L	Occlusion	Evacuation, Dacron patch
24	Hamming & Vink[34]	1965	—	—	—	—	—
25	Harris & Jepson[35]	1965	11	M	R	Stenosis	Evacuation
26	Mentha[62]	1965	33	M	R	Stenosis	Aspiration
27	Hart Hansen[36]	1966	56	M	R	Occlusion	Evacuation
28	Descotes, et al.[21]	1966	48	M	L	Stenosis	Evacuation
29	Pierangeli & De Rubertis[71]	1966	39	M	L	Stenosis	Resection, vein graft
30	Barnett, Dugdale & Ferguson[5]	1966	61	M	R	Stenosis	Resection, vein graft
31	Bartos, Kalus & Possner[7]	1966	29	M	L	Occlusion	Resection, vein graft
32	Morino, Silvestrini & Galli[64]	1966	42	M	L	Occlusion	Excision, Dacron patch
33	Lewis, et al.[55]	1967	13	M	L	Stenosis	Evacuation, vein patch
34	Lewis, et al.[55]	1967	42	M	L	Stenosis	Evacuation, 1964 & 1966
35	Lewis, et al.[55]	1967	55	M	R	Stenosis	Evacuation
36	Taylor, Taylor & Ramsay[90]	1967	32	M	R	Stenosis	Evacuation
37	Flanc[29]	1967	33	F	L	Stenosis	Evacuation, vein graft
38	Linquette, et al.[56]	1967	35	M	L	Stenosis	Evacuation
39	Stirling & Aarons[87]	1967	40	M	R	Stenosis	Resection, vein graft
40	Imamura, et al.[43]	1967	28	M	L	Stenosis	Partial excision of cyst
41	Baumann*	1967	53	M	L	Occlusion	Resection, vein graft
42	Derom*	1968	(45)	M	—	Stenosis	Evacuation
43	Hofmann, et al.[41]	1969	26	M	R	Stenosis	Evacuation
44	Hierton*	1969	50	F	R	Stenosis	Resection, vein graft
45	Savage[77]	1969	30	M	L	Occlusion	Evacuation
46	Tracy, Ludbrook & Rundle[91]	1969	39	M	R	Stenosis	Evacuation
47	Tracy, Ludbrook & Rundle[91]	1969	35	M	L	Stenosis	Excision, Teflon® graft
48	Tracy, Ludbrook & Rundle[91]	1969	25	M	R	Stenosis	Excision, end-to-end anastomosis
49	Ehringer, et al.[25]	1969	48	M	R	Occlusion	Excision, vein patch
50	Laurendeau[51]	1969	33	M	R	Occlusion	Evacuation
51	Haid, Conn & Bergan[33]	1970	44	M	L	Occlusion	Excision, vein graft
52	Powis, et al.[72]	1970	35	M	L	Occlusion	Evacuation
53	Lord[58]	1970	30	M	R	Stenosis	Excision, suture
54	Little & Goodman[57]	1970	43	M	R	Stenosis	Evacuation, vein bypass
55	Suy, et al.[89]	1970	43	M	R	Occlusion	Evacuation
56	Suy, et al.[89]	1970	46	M	L	Stenosis	Evacuation
57	Suy, et al.[89]	1970	23	M	R	Stenosis	Evacuation
58	Denck*	1970	27	M	L	Stenosis	Excision, Dacron patch
59	Soots[85]	1970	38	M	R	Stenosis	Resection, vein graft
60	Derom*	1971	(45)	M	—	Stenosis	Patch
61	Milliken[63]	1971	32	F	R	Occlusion	Evacuation
62	Shannon[82]	1971	64	M	R	Stenosis	Evacuation, patch
63	Chandler[15]	1971	43	M	R	Occlusion	Evacuation, vein patch
64	Kugimiya, et al.[49]	1971	56	M	R	Stenosis	Partial resection, end-to-end anastomosis
65	Eastcott*	1971	50	M	R	—	Evacuation

TABLE 1. (*Continued*)

No.	Author	Year	Age	Sex	Side	Arteriogram	Treatment
66	Ruppell, Sperling, et al.[76]	1971	46	M	L	—	Evacuation
67	Savage[78]	1972	38	M	R	Occlusion	Evacuation, Dacron® patch
68	Sperling, Schott & Ruppell[86]	1972	52	F	R	Stenosis	Resection, vein graft
69	Soots*	1972	50	M	L	Occlusion	Resection, vein graft Replaced by Dacron graft
70	Cousin, et al.[18]	1973	13	M	—	Occlusion	Resection, vein graft
71	Shute & Rothnie[83]	1973	43	M	R	Stenosis	Evacuation
72	Shute & Rothnie[83]	1973	55	M	R	Stenosis	Evacuation
73	DeLaurentis, et al.[20]	1973	11	F	L	Stenosis	Resection, vein graft
74	Ohara & Minaguhi[67]	1973	62	F	R	Occlusion	Resection, vein graft
75	Ohara & Minaguhi[67]	1973	32	M	R	Stenosis	Resection
76	Dunant & Eugenidis[22]	1973	27	M	L	Stenosis	Resection, vein graft
77	Zinicola, Ferrero & Odero[94]	1973	70	M	L	Stenosis	Evacuation
78	Zinicola, Ferrero & Odero[94]	1973	35	M	L	Stenosis	Resection, vein graft
79	Forti & Tattoni[30]	1973	—	—	—	—	—
80	Dye & Javid*	1973	44	M	R	Occlusion	Resection, vein graft
81	Muller-W. & Papachrysanthou[66]	1974	50	M	R	Occlusion	Resection
82	Darling*	1974	39	M	R	Stenosis	Evacuation, vein patch‡ Recurrence 18 months Saphenous vein graft
83	Baumann*	1974	44	M	L	Stenosis	Evacuation
84	Baumann*	1974	31	M	R	Occlusion	Evacuation, vein patch
85	Eastcott*	1974	11	F	R	Occlusion	Endarterectomy, vein patch
86	Mateo[60]	1974	48	F	—	Occlusion	Resection, Dacron patch
87	Jurado, et al.[46]	1974	—	—	—	—	—
88	Scobie & Curry[80]	1975	54	M	L	Stenosis	Resection, vein graft
89	Muller & Rodriguez[65]	1975	38	F	—	Stenosis	Evacuation
90	Alm[1]	1975	43	M	R	Stenosis	Resection, vein graft
91	Raithel 9 Hacker[73]	1975	27	M	—	—	Evacuation, vein patch
92–97	Gedeon[31]	1975	(6 cases, no details provided)				
98	Kjaergaard & Svendsen[48]	1976	43	M	L	Occlusion	Endarterectomy
99	Waibel*	1976	37	M	—	Stenosis	Resection, vein graft
100	Blum[11]	1976	34	M	R	Scimitar sign	Evacuation
101	Kairaluoma, Karkola & Larmi[47]	1976	46	M	L	Yes	Evacuation
102	Schlesinger & Gottesman[79]	1976	56	M	L	Occlusion	Evacuation
103	Shabbo[81]	1976	44	M	L	Yes	Resection, Dacron graft
104	Faenza[28]	1976	24	M	R	Occlusion	Resection, vein graft
105	Leu[54]	1977	32	M	—	—	—
106	Leu[54]	1977	58	F	—	—	—
107	Leu[54]	1977	62	F	—	—	—
108	Bollinger & Pouliadis[12]	1977	—	—	R	Crescent	—
109	Bollinger & Pouliadis[12]	1977	—	—	R	Crescent	—
110	Bollinger & Pouliadis[12]	1977	—	—	L	Crescent	—
111	Brunner & Soyka[14]	1977	32	M	R	Crescent	Resection, vein graft
112	Brunner & Soyka[14]	1977	54	F	L	Scimitar sign	Evacuation
113	Brunner & Soyka[14]	1977	62	F	L	Scimitar sign	Evacuation
114	Darling*	1977	61	M	L	—	Saphenous vein bypass
115	Haid*	1977	—	—	—	—	Aspiration

* Personal communication to correspondence office. †Cyst located in a branch of popliteal artery but causing stenosis of the popliteal arterial lumen and necessitating resection of a segment of the popliteal artery itself. ‡Initial operation performed by another surgeon.

anastomosis of the popliteal artery in three instances. Among the nonresectional techniques, open evacuation of the cystic cavity was performed in 54 patients while operative aspiration was done in two. Thirteen of the patients undergoing evacuation required additional arterial repair with vein patch being employed in nine patients and synthetic patch angioplasty in four.

Follow-up of these cases is shown in Tables 2 and 3. The mean follow-up of the nonresectional group of patients was 20 months, with the longest follow-up reaching 16 years. In patients who were treated by resectional therapy, the mean follow-up was 33 months, with a maximum follow-up of 18 years.[37,38]

In order to assess the efficacy of cyst evacuation alone, 41 case reports were identified in which cyst evacuation was the only initial treatment reported (Fig. 3). Four of these 41 patients required additional procedures because of recurrence of the cyst. Three of the four were successfully treated by re-evacuation, while the fourth had only mild claudication as a recur-

TABLE 2. *Follow-up Nonresectional Group*

Procedure	Mean Follow-up	Longest Follow-up
Evacuation	22.7 Months	16 Years
Evacuation with vein patch	18.4 Months	3 Years
Evacuation with synthetic patch	—	—
Aspiration	24 Months	4 Years
Total nonresectional goup	20 Months	16 Years

rent symptom and further therapy was deferred. While there were only nine patients treated by initial cyst evacuation with patch angioplasty using autogenous veins, two did develop patch aneurysms which required repair by resection of the affected vessel and its patch and interposition of autogenous vein. Four patients were initially treated by evacuation of the cyst and angioplasty utilizing synthetic patch material. One of these failed immediately postoperatively, presumably from technical fault, and was successfully reoperated.

There were 30 patients initially treated by resection of the affected vessel and interposition vein grafting (Fig. 4, Table 4). Two of these operations failed. One was treated subsequently by sympathectomy alone. The patient's recurrent symptoms of moderate claudication continued. The second failure was treated by replacement of the occluded vein graft by a Dacron® graft. This graft also occluded six months postoperatively.

Initial treatment of the condition involved resection of the affected vessel and replacement by synthetic graft in seven patients. Only one graft thrombosis was reported. This patient was treated by sympathectomy rather than reoperation upon the affected segment. Arterial resection and end-to-end anastomosis was reported in three instances. Twice homografts were utilized successfully.

Tables 5 and 6 display an analysis of the treatment results according to procedure used. As shown there, evacuation in 45 patients produced a successful result in 87% of patients so treated. Vein patch repair in nine instances was totally successful in seven (78%), while one failure was noted in five synthetic patch repairs (80% successful). Open aspiration in two instances was totally successful.

Historically, treatment of this condition extends to the first days of direct vascular reconstruction. However, only one patient has suffered a grossly unsuccessful repair which eventuated in amputation. This patient was treated by vein graft initially and Dacron graft subsequently. When both grafts failed, he lost his leg to severe ischemia.

As shown in Table 5, in patients treated by nonresectional technique, there were nine failures in 61 procedures for an overall success rate of 85%.

When the results of resectional therapy are analyzed (Table 6), it can be seen that vein graft replacement of the arterial segment was eminently successful. There were two failures in 32 attempts (94% success). Even synthetic grafting in this situation appeared to be satisfactory; two failures in eight instances (75% success). The overall success rate in 45 procedures was 91%.

In the total group of 98 patients (106 procedures), one patient required late amputation for graft failure, and two patients were left with residual claudication. Considering that the treatment in all instances consisted of local procedures upon the popliteal artery itself, this analysis of the results shows a remarkably high degree of success.

An overview of treatments suggests that choice of procedure in an individual case is dependent upon the anatomic situation identified at operation as influenced by preoperative findings. As in other forms of arterial reconstructive surgery, no single procedure can be suggested for application to all cases. The first principle of treatment is that cyst evacuation is effective. Patients best treated by cyst evacuation are those in which the artery is not totally occluded and in whom degeneration of the arterial wall is not present. Intraoperative aspiration, evacuation or excision of the cyst can be performed as necessary to restore normal arterial flow through the untouched lumen of the vessel. Intraoperative documentation of perfect distal arterial flow is a requirement of such nonresectional therapy.

A second principle of treatment is that local angioplasties should be avoided. These include venous patch angioplasty, synthetic patch angioplasty, and excision of the cyst with direct suture of the remaining vessel. None of these procedures reliably cures the condition.

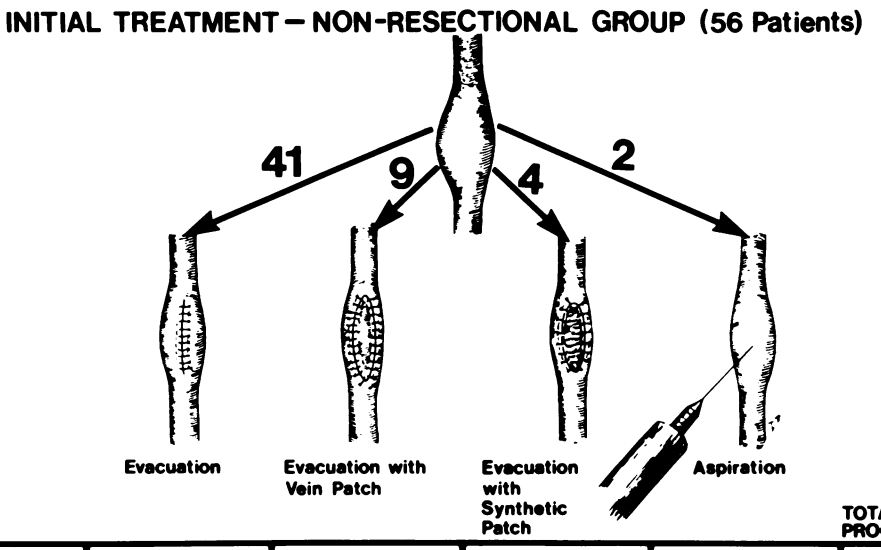
A third principle of treatment is that when total popliteal artery occlusion is present, resection of the lesion or bypass is appropriate. Such replacement or bypass should employ the best techniques of modern vascular surgery including autogenous venous grafting with end-to-side or terminolateral anastomosis to avoid suture line stenosis of the vessel.

Cyst Content

The earliest description of the chemical content of these cysts was from the first report by Ejrup and

TABLE 3. *Follow-up Resectional Group*

Procedure	Mean Follow-up	Longest Follow-up
Vein graft	38 Months	18 Years
Synthetic graft	30 Months	5 Years
Homograft	14.7 Months	1.5 Years
End-to-end anastomosis	12 Months	3 years
Total resectional group	33 Months	18 Years

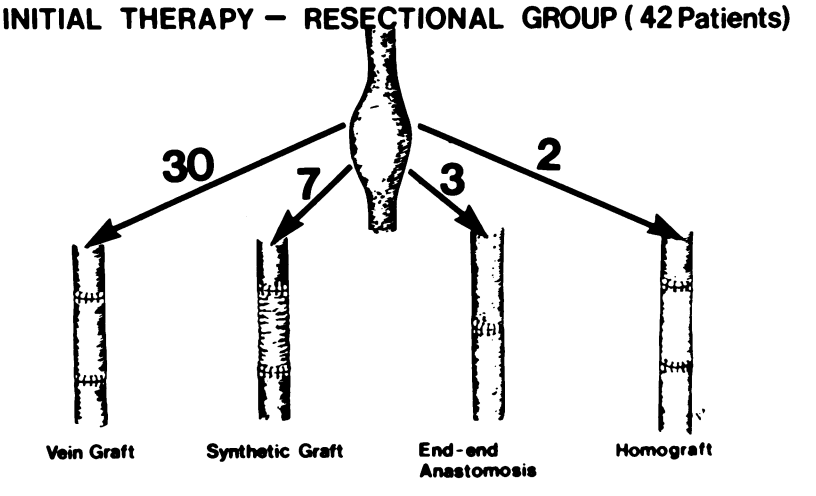


					TOTAL PROCEDURES
FAILURES	4/41	2/9	1/4	0/2	7/56
INITIAL SUCCESS	90%	78%	75%	100%	89%

FIG. 3. Results of initial treatment in nonresected patients with adventitial cystic disease. This pictogram summarizes the results of initial treatment of this condition in those cases in which resection was not done.

Hierton,[26] in which material was shown to contain "abundant fibrinogen, carbohydrate-rich globulin, and hemoglobin." It will be recalled that Hierton had described the gross cyst contents as resembling raspberry jelly. Therefore, the hemoglobin could be a result of hemorrhage into the cyst. Chemical study of the cyst by Dr. B. Swedin showed occasional amino acids, no carbohydrates, no cholesterin or calcium. The conclusion was that hyaluronic acid was the main component. In reviewing this conclusion, Endo[27] pointed out that the method used by Swedin was not specific for hyaluronic acid but could also digest chondroitin sulfate A and C. Endo showed with specific digestion with hyaluronidase from *Streptomyces hyalurolyticus* that the main substance in the cyst obtained by Ohara[67] was proteohyaluronic acid.

Later, Dr. M. L. Welby's analysis of the cyst contents of the case of Harris and Jepson[35] showed "a significant amount of hydroxyprolene suggesting an origin from collagen tissue." However, the analysis performed by Leaf[53] on the cyst contents of case 2 of Lewis et al.[55] failed to detect hydroxyprolene and sug-



					TOTAL PROCEDURES
FAILURES	2/30	1/7	0/3	0/2	3/42
INITIAL SUCCESS	93%	86%	100%	100%	93%

FIG. 4. Results of treatment of adventitial cystic disease of the popliteal artery by resection, indicating the overall excellence of the results.

TABLE 4. *Treatment of Initial Failures*

Initial Procedure	Failures	Reoperation	Success/ Failure	Outcome of Treatment Failure
Evacuation	4/41	Re-evacuation	2/1	Persistent claudication*
		None	0/1	Persistent claudication
Evacuation with vein patch	2/9	Resection with vein graft	2/0	Successful
Evacuation with synthetic graft	1/4	Repatch with synthetic graft	1/0	Successful
Vein graft	2/30	Sympathectomy	0/1	Persistent claudication
		Resection with synthetic graft	0/1	Amputation
Synthetic graft	1/7	Sympathectomy	1/0	Doing well

* This patient developed another recurrence and was successfully treated by repeat evacuation.

gested that the main cyst constituent might be mucoprotein.

Finally, the histochemical characterization of mucin as done by De Laurentis and his group[20] indicates that acid mucopolysaccharides within the cyst are mostly rich in hyaluronic acid radicals, thus supporting the theory that the mucin of the cyst is more likely of ground substance nature, rather than secreted by epithelial cells. These chemical analyses are important in lending weight to the theories of etiology as discussed below.

Theories of Etiology

Clearly, the single theory of etiology that has found the greatest support among observers of the lesion is that of repeated trauma. There is general agreement that there is no evidence whatsoever for suggesting that hemorrhage, neoplasm, nor inflammation could cause the cysts. The original article by Hierton and Lindberg[39] suggested a traumatic etiology and pointed out that early literature on the subject of popliteal artery degeneration by Boyd and Jepson[13] indicated that the popliteal artery enters a fibrous tunnel formed by fascia of the deep surface of the gastrocnemius muscle and is subject to minor trauma or even traumatic thrombosis in this area.

Microtraumatic Origin

Such a theory of microtrauma finds a great number of adherents, including Hoffmann, Consiglio, Hofmeier and Schlosser,[41] Sperling and Ruppell;[86] and Ishikawa,[44] who showed that in patients with cystic disease of the popliteal artery, repeated stretch injuries cause degeneration of the arterial adventitia. A definite traumatic event preceding discovery of the cyst can be identified in a few patients such as Flanc's case,[29] in which a clear traumatic episode occurred falling from a bus; in the case of Tytgat,[92] in which a pedal cycle was implicated; and in that of Holmes,[42] a patient who was subjected to repeated kneeling. However, Savage[78] has summarized the lingering doubts of many observers, stating that, if trauma is a likely cause, "It is difficult to understand why the condition is not more common."

Embryologic Theory

Apart from trauma, the next most popular theory seems to be that of an embryologic origin in which incorporation of mucin secreting cells from the endothelium of the knee joint appear in the adventitia of the artery. These cells, secreting small amounts of mucin over many years, eventually give rise to tense adventitial cysts, which then encroach on the lumen of the vessel. If this were true, the chemical content of the cyst would likely be more characterized by epithelial secretions, rather than by collagen or ground substance breakdown products.

Cysts as Ganglia

Several authorities on the subject feel that the cysts are true ganglia. In the case of Patel and Cormier,[69] a

TABLE 5. *Overall Success Nonresectional Therapy*

Procedure	Failures/ Total Cases	Overall Success Rate
Evacuation	6/45	87%
Evacuation with vein patch	2/9	78%
Evacuation with synthetic graft	1/5	80%
Aspiration	0/2	100%
Total procedures	9/61	85%

TABLE 6. *Overall Success Resectional Therapy*

Procedure	Failures/ Total Cases	Overall Success Rate
Vein graft	2/32	94%
Synthetic graft	2/8	75%
Homograft	0/2	100%
End-to-end anastomosis	0/3	100%
Total procedures	4/45	91%

tendinous ganglion on the posterior surface of the knee was present in addition to a cystic condition of the popliteal artery. No communication existed between these two lesions. Backstrom, Ostberg et al.[4] found cystic mucoid degeneration of a radial artery when they were operating for recurrent ganglion of the wrist, and Parkes has pointed out that similar cystic degeneration of the ulnar artery has been found communicating with the wrist. These observations may not be relevant to the popliteal artery but, in Hart Hansen's description,[36] he suggests that at several operations for cystic mucoid degeneration of the popliteal artery, multiple multilocular cysts have been found in the intramural location of minor arteries arising from the popliteal, and that these cysts have been directed toward adjacent joints.

Darling, in a case reported to this office,* described that the cyst formation was an extension from the adjacent joint capsule. In addition, in this case, ganglion-like fluid was present at an anomalous high origin of the anterior tibial artery. These facts are well illustrated in a drawing by Darling which accompanies his case report.

Shute and Rothnie[83] have also demonstrated communication between the cyst and a neighboring joint but feel that the cellular inclusion theory is supported by this observation.

It has been pointed out that direct communication with the joint is not necessarily important to this theory, since McEvedy[61] suggests that, in ganglia or Baker's cysts in the same location, communication with the joint is not always observed. Blum and Giron[11] support this, saying that traumatic implantation of synovial cells within the adventitia of the artery is a very acceptable explanation. Robb[75] has thought that the cyst formation was from a piece of synovial membrane from tendon sheath or the knee joint itself.

Cystic degeneration of nerves is exceedingly rare and Hansen[36] states categorically that, to the best of his knowledge, cystic degeneration of nerves other than the peroneal has not been described. Clark[17] and Parkes,[68] on the other hand, describe cystic degeneration of the peroneal nerve in the location immediately adjacent to that of the popliteal artery. Parkes† says that the observations in his two cases, one affecting the ulnar artery and one affecting the radial artery, were not influenced by his observations of the intraneural ganglion of the popliteal nerve. He states, "I am certain that I was not influenced by this. The loculated cysts were situated beneath the adventitia of the arteries and there was a definite pedicle continuous with the cyst cavities which led into a neighboring joint. In the case of the radial artery, it was the carpometacarpal joint of the thumb. In both cases, I resected the pedicle and merely decompressed the cysts around the arteries. Neither case has had any recurrence."

Leu[54] from Zurich states categorically that the location of the lesions and the histologic findings indicate that adventitial cysts are true ganglions which originate from adjacent joint capsule or tendon sheath. Brunner,[14] also from Zurich, agrees that the adventitial cysts are ectopic ganglia.

Thus, it seems that the best theory of cyst formation is the latter, in which joint capsular degeneration produces connective tissue change in which cells secrete a substance derived from ground substance or collagen which contains hydroxyprolene. These cells form cysts which invade adventitia. Later, as multiple cysts thus form, enlarge and coalesce with one another to form multilocular cavities, a sudden rupture of one cavity into another can produce rapid growth and encroachment on the arterial lumen. This would explain the sudden onset of symptoms which occurs in most patients. Later, frank necrosis of the media is caused by direct compression between the cyst itself and the thrombus which forms within the arterial lumen.

There is almost no support for congenital or developmental systemic abnormality, although in Linquette's article,[56] a skin biopsy was done which showed changes in the elastic tissue of the skin. This is the only report in which a constitutional vascular fragility was thought to be important in the causation of this condition.

Conclusions

The lesion under discussion remains a rare and fascinating vascular occurrence. It has been possible to identify more than 100 such cases and to note the peculiar grouping of these upon the European continent and in Australasia. Why so few cases are reported from the North American continent remains an item of conjecture. That the treatment of this condition is attended by such a high degree of success is a tribute to the skill of surgeons working at times when vascular surgery was indeed exploratory. Perhaps it is also a tribute to the patients who bring to the problem an excellent inflow and outflow tract to be reconstructed. While the cause of the lesion is undetermined, its relationship to true ganglia is difficult to question. Further observations by operating surgeons in the future should provide valuable information regarding the etiology of this condition. Careful dissection of the cysts, noting attachment and possible communication to adjacent joints, would be of inestimable value. Further analysis of cyst contents with comparison to contents of ganglia might also prove to be a fruitful study.

* Personal communication to Correspondence Office.
† Personal communication to Correspondence Office.

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