Adventitial Cystic Disease (Of The Popliteal Artery): Pathogenesis, Symptoms, And Optimal Treatment .

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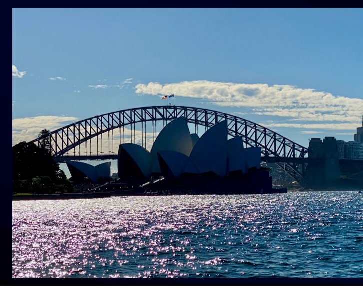
I have no disclosures



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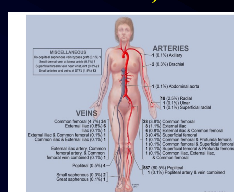
History of Adventitial Cystic Disease (or Cystic Adventitial Disease, ACD or CAD)

VEITH SYMPOSIUM

- First described in 1947 involving the right external iliac artery,
- Around 600 reports have been published since then.
- The popliteal artery is affected in >80% of cases,
- Other vascular sites including veins have been reported.



. Artistic drawing demonstrating the various sites of cystic adventitial disease (CAD) reported in the human body

Atkins H, Key J. A case of myxomatous tumor arising in the adventitia of the left external iliac artery. Br J Surg 1947;34:165-75.
Desy NM, Spinner RJ, The etiology and management of cystic adventitial disease, Journal of Vascular Surgery, 2014; 60(1): 235-245.

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Demographics

VEITH SYMPOSIUM

- Arterial ACD predominantly affects males (M:F=4:1).
- Venous ACD has a male to female ratio of 1 to 1.6:1.
- typical age of presentation is mid-40s.
- The prevalence of cystic adventitial disease is found to be 1:1200 cases of claudication.
- The etiology of ACD remains a matter of controversy and several theories have been proposed.

FlaniganDP,BurnhamSJ,GoodreauJJ,BerganJJ.Summaryofcasesof adventitial cystic disease of the popliteal artery. Ann Surg 1979;189: 165-75.
Vasudevan A, Halak M, Lee S, Ong S, Nadkarni S. Cystic adventitial disease: a case report and literature review. ANZ J Surg 2005;75: 1120-2.
Levien JL, Benn CA. Adventitial cystic disease: A unifying Hypothesis. J Vasc Surg 1998;28:193-205.
Desy NM, Spinner RJ, The etiology and management of cystic adventitial disease, Journal of Vascular Surgery, 2014; 60(1): 235-245

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Theories of Pathogenesis of ACD

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- **The trauma theory:** a chronic degeneration caused by repetitive trauma
- **The ganglion theory:** synovial cysts enlarge and track along vascular branches to eventually implant in the adventitia of adjacent major vessels.
- **The systemic disorder theory:** degeneration and cyst formation of the adventitial layer occurs as part of a generalized connective tissue disorder
- **The developmental theory:** mucin secreting mesenchymal cells from nearby joints are in the adventitia of vessels during the embryogenesis and mucin secretion leads to cyst formation.

FlaniganDP,BurnhamSJ,GoodreauJJ,BerganJJ.Summaryofcasesof adventitial cystic disease of the popliteal artery. Ann Surg 1979;189: 165-75.
Vasudevan A, Halak M, Lee S, Ong S, Nadkarni S. Cystic adventitial disease: a case report and literature review. ANZ J Surg 2005;75: 1120-2.
Levien JL, Benn CA. Adventitial cystic disease: A unifying Hypothesis. J Vasc Surg 1998;28:193-205.

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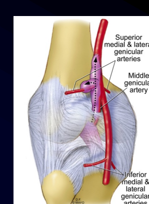
Developmental Theory

VEITH SYMPOSIUM

- **Connections between the adventitial cyst and the capsule of the adjacent joint have been found on imaging and identified intraoperatively in up to 17% of cases, supporting some form of developmental anomaly.**
- **A modified developmental theory suggested that less differentiated joint-related mesenchymal cell rests are responsible for cystic adventitial disease.**
- **Simultaneous development of both nonaxial arteries and joints supports the embryologic hypothesis that mesenchymal tissue is entrapped in the nearby developing non- axial vessels.**
- **Mesenchymal cell rests in the arteries lead to the formation of a cystic lesion occurs later in life when these entrapped cells start to secrete mucoid material.**



The articular (synovial) theory for cystic adventitial disease (CAD) formation within the popliteal artery.

Levien JL, Benn CA. Adventitial cystic disease: A unifying Hypothesis. J Vasc Surg 1998;28:193-205.
Spinner RJ, Desy NM, Agarwal G, Pawlina W, Kalra M, Amrami KK. Evidence to support that adventitial cysts, analogous to intraneural ganglion cysts, are also joint-connected. Clin Anat 2013;26:267-81.
Chiche L, Barranger B, Cordoliani YS, Darrieus H, Guyon P, Vicq P. Two cases of cystic adventitial disease of the popliteal artery. Current diagnostic approach. J Mal Vasc 1994;19:57-61.
GalleC,CavenaileJC,HoangAD,MinhTL,PenyMO,BraudeP,etal. Adventitial cystic disease of the popliteal artery communicating with the knee joint. A case report. J Vasc Surg 1998;28:738-41
Buijsrogge MP, van der Meij S, Korte JH, Fritschy WM. "Intermittent claudication intermittence" as a manifestation of adventitial cystic dis- ease communicating with the knee joint. Ann Vasc Surg 2006;20: 687-9.
VanhoenackerFM,VandevenneJE,DeSchepperAM,DeLeersnijderJ. Regarding "Adventitial cystic disease: a unifying hypothesis." J Vasc Surg 2000;31:621-2.

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Clinical Presentation

- **Typically affects popliteal artery (over 80% cases) and symptoms of claudication that is usually of short distance or sudden onset or after an episode of exertion.**
- Unlike typical claudication the symptoms may completely resolve for a period of time, and re-present later or progress rapidly.
- **Recovery time from pain is prolonged compared with the typical claudicants.**
- May present with pain behind the knee due to pressure effect
- Acute calf pain on a background of long-standing exertional pain.
- A soft tissue or pulsatile mass.

Devereux D, Forrest H, McLeod T, Ahweng A. The nonarterial origin of cystic adventitial disease of the popliteal artery in two patients. Surgery. 1980;88(5):723-727.
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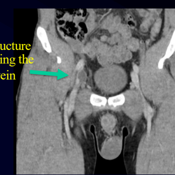
Miller A, Salenius JP, Sacks BA, Gupta SK, Shoukimas GM. Noninvasive vascular imaging in the diagnosis and treatment of adventitial cystic disease of the popliteal artery. J Vasc Surg. 1997;26(4):715-720.
Hunt BP, Harrington MG, Goode JJ, Galloway JM. Cystic adven- titial disease of the popliteal artery. Br J Surg. 1980;67(11): 811-812.
Velasquez G, Zollikofer C, Nath HP, et al. Cystic arterial adven- titial degeneration. Radiology. 1980;134(1):19-21.
Cassar K,Engeset J.Cystic adventitial disease: a trap for the unwary. Eur J Vasc Endovasc Surg. 2005;29(1):93-96.
Maged IM, Kron IL, Hagspiel KD. Recurrent cystic adventitial disease of the popliteal artery: successful treatment with percuta- neous transluminal angioplasty. Vasc Endovascular Surg. 2009; 43(4):399-402.
Pursell R, Torrie EP, Gibson M, Galland RB. Spontaneous and permanent resolution of cystic adventitial disease of the popliteal artery. J R Soc Med. 2004;97(2):77-78.
Peterson JJ, Kransdorf MJ, Bancroft LW, Murphey MD. Imaging characteristics of cystic adventitial disease of the peripheral arteries: presentation as soft-tissue masses. AJR Am J Roentgenol. 2003;180(3):621-625.
Flanigan DP, Burnham SJ, Goodreau JJ, Bergan JJ. Summary of cases of adventitial cystic disease of the popliteal artery. Ann Surg. 1979;189(2):165-175

Presentation

- **Cystic adventitial disease affecting veins leads to limb swelling (due to partial/complete occlusion).**
- **Patients may present with deep vein thrombosis, when the vein is totally occluded.**



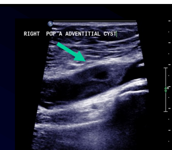
Johnson JM, Kiankhooy A, Bertges DJ, Morris CS. Percutaneous image-guided aspiration and sclerosis of adventitial cystic disease of the femoral vein. Cardiovasc Intervent Radiol. 2009;32(4):812-816.
DixFP,McDonaldM,ObomighieJ,etal. Cystic adventitial disease of the femoral vein presenting as deep vein thrombosis: a case report and review of the literature. J Vasc Surg. 2006;44(4):871-874.

Clinical Assessment

- **Detecting ACD clinically is difficult and requires a high index of suspicion, absence of risk factors.**
- **Age of the patient, Site of lesion, and presenting history, as patients may have a sudden onset of symptoms or rapid progression of claudication symptoms.**
- **Ishikawa sign: Easily palpable distal pulses, that disappear when the knee is flexed, due to the complete occlusion of the vessel during the manoeuvre.**
- **Ankle–brachial pressure index and exercise test may demonstrate a drop in pressure.**
- **Venous CAD clinically is much more difficult, as patients usually present with leg swelling that can easily be diagnosed as deep vein thrombosis.**

Investigations

- Ultrasound
  - **Most adventitial cysts will appear anechoic, perhaps intravascular, and predominantly have the same appearance as simple fluid.**
  - **Appearance of these cysts depends on the mucin content (greater echo- density).**
  - **Low echoic areas may be missed**
  - **Can be mistaken for an aneurysm.**
- IVUS
  - **Has a higher frequency than transcutaneous ultrasound (20 MHz versus 10 MHz) and differentiates circumferentially between the media and adventitia, with little plaque and a normal intima.**
  - **IVUS can also evaluate and differentiate abnormalities of the lumen or vessel wall (thrombosis, PAD, and dissection) , as well as extra-vascular abnormalities (popliteal ganglion, Baker's cyst, and popliteal artery compression syndrome).**



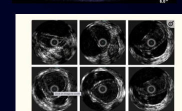


Figure 2

Intravascular ultrasound revealed the nonatherosclerotic lesion and the extrinsic compression of the lumen by the extravascular cyst. The cystic appearance (extravascular hypodensity area) can be visualized clearly.

Niizeki T, Ishino M, Kitahara T, Yamauchi S, Ikeno E, Kubota I. A Case of Cystic Adventitial Degeneration of the Left Popliteal Artery Diagnosed by Intravascular Ultrasound. Clin Med Insights Case Rep. 2016 Feb 29;9:11-4

Investigations

- CT Angiography
  - **CT-Angiography allows three-dimensional reconstructions to demonstrate the cystic nature of the lesion and vessel occlusion.**
- MRI/MRA
  - **Noninvasive, give reproducible results, and most importantly, is useful at demonstrating connections between adventitial cysts and the adjacent joint capsule. The non-angiographic sequences typically show homogenous low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Steady state MRA imaging is much more useful in cases of venous CAD, which is associated with limb edema and where conventional venography may not be reliable.**

Fig. 1. Preintervention Computed Tomography Angiography (CTA) of the popliteal stenosis caused by the cyst. **(A)** Longitudinal reconstruction, **(B)** three-dimensional volume rendering, and axial reconstruction **(C–F)**.

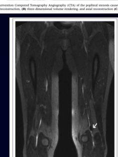
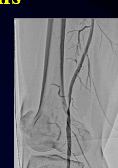


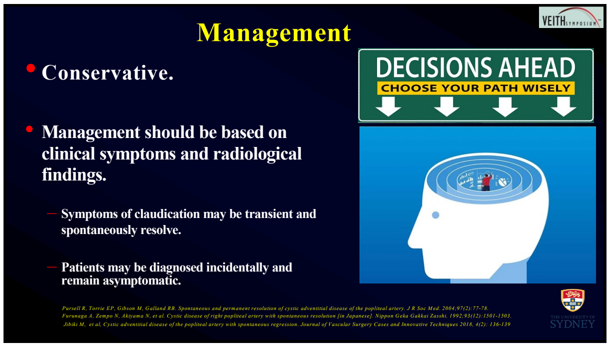
Figure 2. Three-dimensional dynamic magnetic resonance angiography demonstrating a constant filling defect in the proximal popliteal artery, which shows well-circumscribed border in keeping with external compression.

Vento V et al, Cystic Adventitial Disease of the Popliteal Artery: Radical Surgical Treatment After Several Failed Approaches. A Case Report and Review of the Literature . Ann Vasc Surg 2020; 64: 411.e5–411.e11
Paravastu SC, Regi JM, Turner DR, Gaines PA. A contemporary review of cystic adventitial disease. Vasc Endovascular Surg. 2012 Jan;46(1):5-14.

Investigations

- Angiography
  - **Diagnosis of CAD was made based on arteriography. The classic appearance was described as "Scimitar sign," which shows a curvilinear smooth narrowing of the vessel. An "hourglass" configuration may be noted in cases of circumferential lesions.**
  - **Replaced as the primary diagnostic modality by other angiographic techniques (CTA and MRA) that have the capacity to show not only the lumen but also the lesion within the vessel wall**
  - **Conventional venography has been shown to be useful in demonstrating the classical smooth luminal compression suggestive of CAD.**




Management
Conservative.
Management should be based on clinical symptoms and radiological findings.
Symptoms of claudication may be transient and spontaneously resolve.
Patients may be diagnosed incidentally and remain asymptomatic.
DECISIONS AHEAD
CHOOSE YOUR PATH WISELY
Pursell R, Torrie EP, Gibson M, Galland RB. Spontaneous and permanent resolution of cystic adventitial disease of the popliteal artery. J R Soc Med. 2004;97(2):77-78.
Furunaga A, Zempo N, Akiyama N, et al. Cystic disease of right popliteal artery with spontaneous resolution [in Japanese]. Nippon Geka Gakkai Zasshi. 1992;93(12):1501-1503.
Jibiki M, et al, Cystic adventitial disease of the popliteal artery with spontaneous regression. Journal of Vascular Surgery Cases and Innovative Techniques 2018, 4(2): 136-139
VEITH SYMPOSIUM
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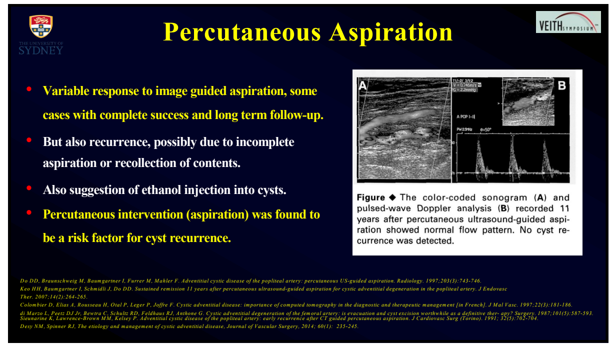
Percutaneous Aspiration
Variable response to image guided aspiration, some cases with complete success and long term follow-up.
But also recurrence, possibly due to incomplete aspiration or recollection of contents.
Also suggestion of ethanol injection into cysts.
Percutaneous intervention (aspiration) was found to be a risk factor for cyst recurrence.
Figure ◆ The color-coded sonogram (A) and pulsed-wave Doppler analysis (B) recorded 11 years after percutaneous ultrasound-guided aspiration showed normal flow pattern. No cyst recurrence was detected.
Do DD, Braunschweig M, Baumgartner I, Furrer M, Mahler F. Adventitial cystic disease of the popliteal artery: percutaneous US-guided aspiration. Radiology. 1997;203(3):743-746.
Keo HH, Baumgartner I, Schmidli J, Do DD. Sustained remission 11 years after percutaneous ultrasound-guided aspiration for cystic adventitial degeneration in the popliteal artery. J Endovasc Ther. 2007;14(2):264-265.
Colombier D, Elias A, Rousseau H, Otal P, Leger P, Joffre F. Cystic adventitial disease: importance of computed tomography in the diagnostic and therapeutic management [in French]. J Mal Vasc. 1997;22(3):181-186.
di Marzo L, Peetz DJ Jr, Bewtra C, Schultz RD, Feldhaus RJ, Anthone G. Cystic adventitial degeneration of the femoral artery: is evacuation and cyst excision worthwhile as a definitive ther- apy? Surgery. 1987;101(5):587-593.
Sieunarine K, Lawrence-Brown MM, Kelsey P. Adventitial cystic disease of the popliteal artery: early recurrence after CT guided percutaneous aspiration. J Cardiovasc Surg (Torino). 1991; 32(5):702-704.
Desy NM, Spinner RJ, The etiology and management of cystic adventitial disease, Journal of Vascular Surgery, 2014; 60(1): 235-245.
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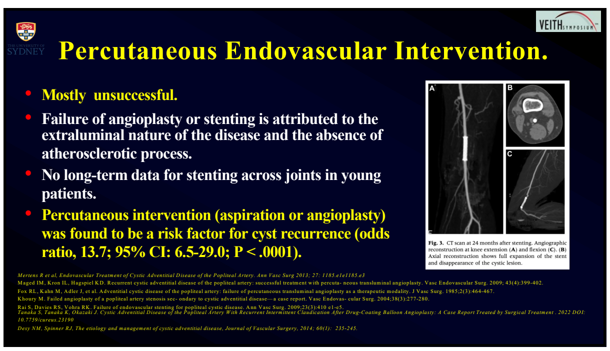
Percutaneous Endovascular Intervention.
Mostly unsuccessful.
Failure of angioplasty or stenting is attributed to the extraluminal nature of the disease and the absence of atherosclerotic process.
No long-term data for stenting across joints in young patients.
Percutaneous intervention (aspiration or angioplasty) was found to be a risk factor for cyst recurrence (odds ratio, 13.7; 95% CI: 6.5-29.0; P < .0001).
Fig. 3. CT scan at 24 months after stenting. Angiographic reconstruction at knee extension (A) and flexion (C). (B) Axial reconstruction shows full expansion of the stent and disappearance of the cystic lesion.
Mertens R et al, Endovascular Treatment of Cystic Adventitial Disease of the Popliteal Artery. Ann Vasc Surg 2013; 27: 1185.e1e1185.e3
Maged IM, Kron IL, Hagspiel KD. Recurrent cystic adventitial disease of the popliteal artery: successful treatment with percuta- neous transluminal angioplasty. Vasc Endovascular Surg. 2009; 43(4):399-402.
Fox RL, Kahn M, Adler J, et al. Adventitial cystic disease of the popliteal artery: failure of percutaneous transluminal angioplasty as a therapeutic modality. J Vasc Surg. 1985;2(3):464-467.
Khoury M. Failed angioplasty of a popliteal artery stenosis sec- ondary to cystic adventitial disease—a case report. Vasc Endovas- cular Surg. 2004;38(3):277-280.
Rai S, Davies RS, Vohra RK. Failure of endovascular stenting for popliteal cystic disease. Ann Vasc Surg. 2009;23(3):410 e1-e5.
Tanaka S, Tanaka K, Okazaki J. Cystic Adventitial Disease of the Popliteal Artery With Recurrent Intermittent Claudication After Drug-Coating Balloon Angioplasty: A Case Report Treated by Surgical Treatment . 2022 DOI: 10.7759/cureus.23190
Desy NM, Spinner RJ, The etiology and management of cystic adventitial disease, Journal of Vascular Surgery, 2014; 60(1): 235-245.
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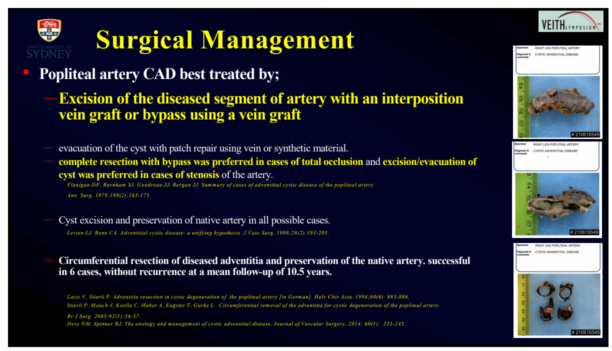
Surgical Management
Popliteal artery CAD best treated by;
Excision of the diseased segment of artery with an interposition vein graft or bypass using a vein graft
evacuation of the cyst with patch repair using vein or synthetic material.
complete resection with bypass was preferred in cases of total occlusion and excision/evacuation of cyst was preferred in cases of stenosis of the artery.
Flanigan DP, Burnham SJ, Goodreau JJ, Bergan JJ. Summary of cases of adventitial cystic disease of the popliteal artery. Ann Surg. 1979;189(2):165-175.
Cyst excision and preservation of native artery in all possible cases.
Levien LJ, Benn CA. Adventitial cystic disease: a unifying hypothesis. J Vasc Surg. 1998;28(2):193-205.
Circumferential resection of diseased adventitia and preservation of the native artery. successful in 6 cases, without recurrence at a mean follow-up of 10.5 years.
Lazic V, Stierli P. Adventitia resection in cystic degeneration of the popliteal artery [in German]. Helv Chir Acta. 1994;60(6): 883-886.
Stierli P, Mauch J, Koella C, Huber A, Eugster T, Gurke L. Circumferential removal of the adventitia for cystic degeneration of the popliteal artery. Br J Surg. 2005;92(1):56-57.
Desy NM, Spinner RJ, The etiology and management of cystic adventitial disease, Journal of Vascular Surgery, 2014; 60(1): 235-245.
RIGHT LEG POPLITEAL ARTERY
CYSTIC ADVENTITIAL DISEASE
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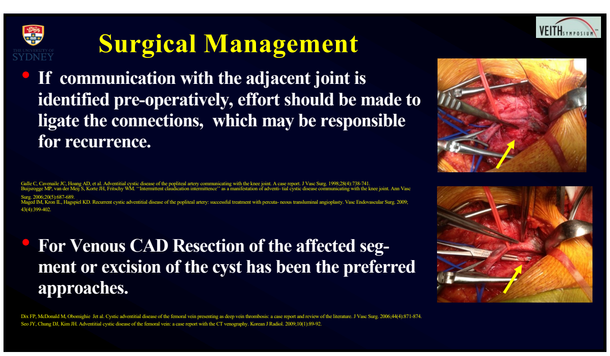
Surgical Management
If communication with the adjacent joint is identified pre-operatively, effort should be made to ligate the connections, which may be responsible for recurrence.
Galle C, Cavenaile JC, Hoang AD, et al. Adventitial cystic disease of the popliteal artery communicating with the knee joint. A case report. J Vasc Surg. 1998;28(4):738-741.
Buijsrogge MP, van der Meij S, Korte JH, Fritschy WM. ''Intermittent claudication intermittence'' as a manifestation of adventi- tial cystic disease communicating with the knee joint. Ann Vasc Surg. 2006;20(5):687-689.
Maged IM, Kron IL, Hagspiel KD. Recurrent cystic adventitial disease of the popliteal artery: successful treatment with percuta- neous transluminal angioplasty. Vasc Endovascular Surg. 2009; 43(4):399-402.
For Venous CAD Resection of the affected seg- ment or excision of the cyst has been the preferred approaches.
Dix FP, McDonald M, Obomighie Jet al. Cystic adventitial disease of the femoral vein presenting as deep vein thrombosis: a case report and review of the literature. J Vasc Surg. 2006;44(4):871-874.
Seo JY, Chung DJ, Kim JH. Adventitial cystic disease of the femoral vein: a case report with the CT venography. Korean J Radiol. 2009;10(1):89-92.
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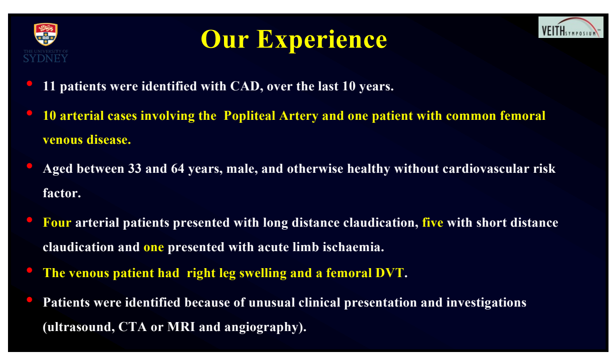
Our Experience
11 patients were identified with CAD, over the last 10 years.
10 arterial cases involving the Popliteal Artery and one patient with common femoral venous disease.
Aged between 33 and 64 years, male, and otherwise healthy without cardiovascular risk factor.
Four arterial patients presented with long distance claudication, five with short distance claudication and one presented with acute limb ischaemia.
The venous patient had right leg swelling and a femoral DVT.
Patients were identified because of unusual clinical presentation and investigations (ultrasound, CTA or MRI and angiography).
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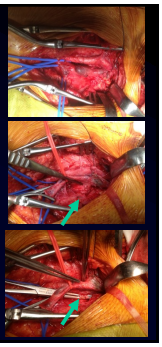
Our Experience

- Arterial patients had local resection and interposition vein grafts
- The median follow up was 50.5 (range 26 to 113) months.
- Nine (9) patients did well with normal post-operative ABI.
- 1 patient developed recurrence at the surgical site and underwent local re-excision and redo bypass.
- The patient with venous ACD presented with a right common femoral vein obstruction. He was treated with resection of the diseased venous segment, and interposition vein grafting.



intraoperative photos of the femoral vein with ACD, and a sinus communicating posteriorly to hip joint and ligated

Thank you