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The Pulse on Stroke in Pulseless Disease (Takayasu Arteritis)

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Takayasu arteritis, first described during an ophthalmology meeting in 1908, is a rare and chronic systemic vasculitis of unknown mechanism mainly involving the aorta, its major branches, and the retinal arteries.[1, 2] Pathologically, it is a pan-arteritis with fibrous thickening and destruction of all layers of the artery, and cellular infiltration around the vasa vasorum. Young women are predominantly affected. The disease is more prevalent in Asia (especially Japan) and Latin America as compared to Western countries. Obstruction of the subclavian or brachial arteries frequently results in loss of the radial pulse (hence the name 'pulseless disease'). Ischemic symptoms in the brain, retina, and other organs result from arterial stenosis or thrombus formation, and rarely dissection or aneurysmal rupture.

Prior studies have shown a high prevalence of stroke in Takayasu arteritis, and a substantial proportion develop stroke as its first manifestation.[3, 4] Knowledge about stroke presentations, risk factors, recurrence, and prevention in Takayasu arteritis has been lacking. To address this, the French Takayasu Network conducted a retrospective analysis of 320 patients with Takayasu arteritis encountered over nearly 45 years (1970–2014) in multiple referral centers.[5] Strengths of the study include the large number of patients with this rare disease, the use of accepted diagnostic criteria, the detailed pictorial depiction of affected arteries and stroke territories involved, treatment information, and long follow-up. In this study 13% developed ischemic stroke and 7% developed TIA at the time of diagnosis or during follow-up. This may be an under-estimate since 75% of stroke/TIA patients were diagnosed after the year 2000 and their diagnosis was based on imaging which has evolved and not available in the initial study years. Stroke/TIA occurred before the eventual diagnosis of Takayasu arteritis in 60%, showing how important it is palpate arterial pulses, look for blood pressure differences between arms, do a funduscopic examination, and consider tests for vasculitis in young individuals with stroke.

The vast majority of stroke/TIA involved the anterior circulation. No patient developed brain hemorrhage. Interestingly, carotidynia symptoms and prolonged time to diagnosis, but not intuitive risk factors such as supra-aortic involvement, degree of stenosis or intracranial artery involvement, or atherosclerosis risk factors, were associated with stroke/TIA. Hence the authors postulate that alternative mechanisms such as microvascular involvement may also contribute to cerebrovascular events.[6] Further studies utilizing perfusion-MRI and

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PET scans are warranted to fully understand mechanisms of stroke in Takayasu arteritis. Of course, the study was likely under-powered, and the results are likely confounded by the more frequent use of stroke preventive medications such as aspirin, clopidogrel and anticoagulants in patients considered 'high risk' due to widespread arterial involvement, and the inclusion of TIA (a less reliable outcome versus imaging-confirmed stroke) in the analysis.

Over half the patients had recurrent stroke. It is sobering that there was no difference in stroke-free survival between patients diagnosed before and after 2000 despite medical advancements. Delayed diagnosis increased the risk for stroke recurrence, emphasizing the ongoing challenges in diagnosing cerebral arteriopathies.[7] While this study provides important information about stroke in Takayasu arteritis, prospective multicenter collaborative studies—similar to ongoing efforts concerning stroke in young adults and rare causes of stroke such as primary CNS vasculitis—are urgently needed to fully understand its mechanisms, risk factors, and develop evidence-based treatment strategies.[8, 9] After all, though uncommon, Takayasu arteritis affects young patients where the occurrence of stroke and even TIA carries a disproportionately high socio-economic and psychological impact.[10–12]

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References

1. Numano F The story of Takayasu arteritis. Rheumatology (Oxford). 2002;41:103–106 [PubMed: 11792888]
2. Agueda AF, Monti S, Luqmani RA, Buttgereit F, Cid M, Dasgupta B, et al. Management of Takayasu arteritis: A systematic literature review informing the 2018 update of the EULAR recommendation for the management of large vessel vasculitis. RMD Open. 2019;5:e001020 [PubMed: 31673416]
3. Couture P, Chazal T, Rosso C, Haroche J, Leger A, Hervier B, et al. Cerebrovascular events in Takayasu arteritis: A multicenter case-controlled study. J Neurol. 2018;265:757–763 [PubMed: 29392458]
4. Duarte MM, Geraldes R, Sousa R, Alarcao J, Costa J. Stroke and transient ischemic attack in Takayasu's arteritis: A systematic review and meta-analysis. J Stroke Cerebrovasc Dis. 2016;25:781–791 [PubMed: 26775269]
5. Mirouse A Ischemic neurological events in Takayasu arteritis patients. Stroke. 2021;XXXX
6. Noel N, Butel N, Le Hoang P, Koskas F, Costedoat-Chalumeau N, Wechsler B, et al. Small vessel involvement in Takayasu's arteritis. Autoimmun Rev. 2013;12:355–362 [PubMed: 22691438]
7. Singhal AB. Diagnostic challenges in RCVS, PACNS, and other cerebral arteriopathies. Cephalalgia. 2011;31:1067–1070 [PubMed: 21673004]
8. Putaala J, Martinez-Majander N, Saeed S, Yesilot N, Jakala P, Nerg O, et al. Searching for explanations for cryptogenic stroke in the young: Revealing the triggers, causes, and outcome (SECRETO): Rationale and design. Eur Stroke J. 2017;2:116–125 [PubMed: 31008307]
9. de Boysson H, Arquizan C, Touze E, Zuber M, Boulouis G, Naggara O, et al. Treatment and long-term outcomes of primary central nervous system vasculitis. Stroke. 2018;49:1946–1952 [PubMed: 29986936]

10. Singhal AB, Biller J, Elkind MS, Fullerton HJ, Jauch EC, Kittner SJ, et al. Recognition and management of stroke in young adults and adolescents. Neurology. 2013;81:1089–1097 [PubMed: 23946297]

11. Singhal AB, Lo W. Life after stroke: Beyond medications. Neurology. 2014;83:1128–1129 [PubMed: 25128181]

12. Maaijwee NA, Rutten-Jacobs LC, Arntz RM, Schaapsmeerders P, Schoonderwaldt HC, van Dijk EJ, et al. Long-term increased risk of unemployment after young stroke: A long-term follow-up study. Neurology. 2014;83:1132–1138 [PubMed: 25128177]