

**Takayasu's Arteritis  
VS  
Aortic Arch Syndrome**

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It has generally been accepted and upheld that, in order to establish a distinct disease entity with absolute validity, at least four conditions must be fulfilled: (1) The distinguishing clinical features which must be characteristic, constant and unequivocal. (2) The pathogenesis or etiologic mechanism of the disease process must be clearly demonstrated and understood. (3) The pathologic features must be unique and specific regardless of wherever the site of involvement is located. (4) The absolute etiologic factor or factors must be definitely established.

Regretfully, there are unaccountable medical nosologic nomenclatures being created or in use in the literature at present which are clearly misnomers and/or unscientific. Looking from semantic point of view, especially for concerned individual interest, this is impressive, of course, if one discards facts and logic. It is, indeed, a wonder why such and such names, which serve merely to confuse the already confused state, keep appearing on the pages of medical publications, a highly regarded scientific material.

It is the purpose of this article to bring up criticism on one of the most discussed entity of our time, the so-called Aortic Arch Syndrome. The article is personal, and is expressed merely as an attempt to clarify the controversial nomenclatures of this condition for academic interest.

Historically, in 1908, a certain Japanese ophthalmologist by the name of Takayasu (Takayashu by some authors) reported a peculiar illness in a Japanese woman to the Japanese Society of Ophthalmologist. This woman, of which no age

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was stated, had gradually developed loss of vision associated with an unknown, wreath-like ocular anastomosis. According to Judge and associates (1962), Takayasu's own case did not have a clinical finding of diminish or absent pulsation. However, upon his case presentation, a discussion which followed, disclosed another case referred by Onishi to have a similar clinical manifestation but, in addition to ocular vascular abnormalities, was also found to be "pulseless". Why then, this condition is not called Onishi's Arteritis rather than Takayasu's is out of the scope of this article.

Since the original case had been published, numerous subsequent cases were reported from Japan and, much later on, from other countries as well. Actually, up to the year 1951, all of the 58 consecutive cases appeared in the literature were homogeneously Japanese. The first case of this disease in North America was reported by Caccamise and Whitman (1952) and was thought to be the first recorded occidental case. In reviewing of the literature and a study from his own two cases, Ask-Upmark (1954) collected a total of 28 occidental patients. His review and study is extensively and excellently done in his publication. From that time on, there were many extra-japaneses cases reported from the world over, both the occidental and the oriental. Concurrently with the piling up of the reports, there is also a piling up of variety of different titles created to describe this same condition. The synonyms are enormous. To cite a few, there are "Pulseless disease" or syndrome, "Aortic Arch Syndrome", Branchial or Brachiocephalic Arteritis "Young female Arteritis", "Marterell's Syndrome" ecetera, ecetera. Among this havoc of names, the Aortic Arch Syndrome appears to enjoy itself more than the others.

The following observation is obtained from review of the English written literature. Attention is given to geographic and sex distribution, clinical pictures, laboratory finding and specifically, the morphopathologic features of the disease. The result of the review is grouped into three headings and criticism is carried out accordingly.

- A: The classical Takayasu's Arteritis.
- B: The so-called Aortic Arch Syndrome
- C: The unnecessary synonyms.

## A : The classical Takayasu's Arteritis

According to the number of cases recorded in the literature, the disease is more prevalent in the orient especially Japan than in other part of the world, although there are cases of other races as well. Among occidental cases, it appears that the Northern European are more affected than the American, also as judged by the number of cases documented in the literature. Clinically, the patient is a young female below 40 years of age. In Japanese series of 44 cases which the ages were recorded, 36 patients were below the age of 25 years old, while in occidental series, 15 out of 28 were below the age of 25. (Shimizu and Sano, 1951; Caccamise and Whitman, 1952; Ask-Upmark, 1954) of the total of 73 oriental cases review by this author, there were only 8 cases which the patients were male (3 Japanese, 4 Chinese and 1 Hindu). The patient usually has a thin wasted face that makes her look older than her age and has a tendency to walk with her head bent forward. (Shimizu and Sano, 1951; Caccamise and Whitman, 1952; Ask-Upmark, 1954; Harrison, 1960; Hirsch and Associates, 1964).

The typical cases should not have a history of any traceable chronic disease with demonstrable relationship to Takayasu's Arteritis. Most characteristic clinical findings besides diminish pulsation of the affected artery, are ocular abnormalities such as vascular anastomosis, visual disturbances, cataracts, corneal opacities and even blindness. (Shimizu and Sano, 1951; Rosssand McKusick, 1952; Judge and Associates, 1962). Signs of central nervous system involvement especially headache, dizziness, convulsions are also common. The disease runs a chronic and periodic remission course.

The laboratory finding show a constant high erythrocytic sedimentation rate and abnormal protein electrophoresis. The serological tests for syphilis, tuberculosis, collagen diseases and other specific tests for specific diseases are not diagnostic. Hirsch and associates had plausibly attempted variety of tests on five cases and none was found to be specific. (Shimizu and Sano, 1951; Ask-Upmark and Fajers, 1956; Wickbom, 1957; Harrison, 1960; Judge and Associates, 1962; Hirsch and Associates, 1964)

The arteriographic finding had been first mentioned by Ask-Upmark and Fajers (1956) and later on, Wickbom, (1957) had stressed the characteristic changes

composed of regularity of the wall at the site of constriction as well as with the occluded region.

The site of involvement, in most cases, is in the brachiocephalic region of the aortic arch. However rare as it may be, this is, by no mean, the only location as Ask-Upmark (1964) has reported in this publication. It is unfortunate that those which there was involvement of extra arch region, on satisfactory specific histopathologic pictures available.

Pathologically, the disease begins as a slow progressive periarteritis (Koszewski and Hubbard, 1957) and then spreads to involve all of the arterial coats. Characteristic features in moderately advanced case include thickening of the intima with marked narrowing of the lumen with or without intravascular thrombosis. The adventitia show fibrosis which may spread to involve the adjacent soft tissue. Periarterial infiltration of plasma cells, lymphocytes and macrophages are present especially around the vasa vasorum, but, endarteritis obliterans of the vasa have near been observed. Fibrinoid necrosis is conspicuously absent. Medial atrophy with focal disruption of the elastic fibers occasionally, with areas of necrobiosis are characteristic. The cellular infiltration may be found in the tunica media. Sometime, multinucleated giant cells are also present. Strikingly, eosinophiles have never been observed, not only in the affected wall, but also in the immediate surrounding vicinity. The most constant intimal changes microscopically is the disruption of the elastic lamella and intimal hyalinization. (Shimizu and Sano, 1951; Mengis et al, 1959; Harrison, 1960; Judge and Associates, 1962; Hirsch and Associates, 1964;)

The absolute nature of the disease is not yet known although it seems that an antigen-antibody reaction may be the most probable cause as judged by the type of granulomatous inflammation, the high erythrocytic sedimentation rate, the abnormal protein electrophoresis and clinically the chronic and remission course of the disease. Because of the age and sex which are almost specific, Ask-Upmark (1956) suggested its relation to some type of collagen diseases especially lupus erythematosus. Judge and associates (1962) speculated an autoimmune reaction to the arterial "elastin" as a possible pathogenesis.

## B: The so-called "Aortic Arch Syndrome"

It has been hitherto agreed that Frovig was the first who applied this title to the condition in 1946 although it was Davy who described the lesion earlier (Judge and Associates, 1962 quoted). Ask-Upmark and Fajers, (1956) later on suggested this to denote only the aortic arch involvement of any condition of whatever nature that characterized clinically by obliteration of any great vessels arising from the aortic arch.

In critical review of this subject in the literature, Judge and associates had excellently demonstrated logically that this term, if continue to be exist, should be reserved to describe only a general CLINICAL ENTITY regardless to the etiology. This author is in complete agreement with their view and would even extend it a little further that this name is inappropriate thus, should not be used any more. The reason is that, in a specific sense, a clinical term can not denote a disease entity until the exact etiologic factor can be established. Besides, what is the use of having so many names when the original Takayasu's Arteritis is well recognised the world over. As for other similar clinical conditions which there is involvement of aortic arch great vessels or involvement of any artery at other region of which nature can be traceable or demonstrated, specific scientific terms should be appropriately applied.

In the vast sea of literature concerning Aortic Arch Syndrome, many of the cases reported, the underlying nature of the diseases could be traced or even established. For instance, there were cases of syphilitic arteritis reported under the name of Aortic Arch Syndrome (Segal and Berezowski, 1958), Tuberculosis (Hirsch and Associates, 1964), Atherosclerosis, (Thurlbeck and Currens, 1959), Rheumatic fever, (Sandring and Welin, 1961; Paloheimo et al, 1966); Rheumatoid arthritis (Sandring and Welin, 1961), Idiopathic thrombocytopenia, trauma, thromboangiitis obliterans and other collagen diseases were also described under the same heading of Aortic Arch Syndrome. (Ross and McKusiek, 1953; Segal and Berezowski, 1958; Thurlbeck and Currens, 1959; Sandring and Welin, 1961; Hirsch and Associates, 1964).

It is therefore suggested that, the term Aortic Arch Syndrome, in which the

underlying nature can be traceable or demonstrated, should be discontinued, and a specific clinicopathologic title replaced. Hence, the name "Syphilitic Subclavian Arteritis" should replace Aortic Arch Syndrome having clinicopathologic features of Syphilis. "Atheromatous Innominate Arteritis" should be used in Aortic Arch Syndrome associated with atherosclerosis. "Traumatic common carotid Arteritis" should replace Aortic Arch Syndrome having an established evidence of trauma, and so on and so on. As for Aortic Arch Syndrome which definitely, no etiology could be traced or established, then the disease should be called Takayasu's Arteritis. This term, therefore, would be used and understood by all to represent a disease entity characterized by specific clinicopathologic features of unknown etiology.

C: The unnecessary synonyms.

Numerous titles are being used in the literature to denote this condition. Most of them are misnomers as Ask-Upmark and later on Judge and associates have pointed out in their respective excellent study and review.

"Pulseless disease", is entirely equivocal and unscientific. It should be used by a lay man rather than any scientist. Is arterial pulsation in the entire body can really be felt and pulsless? Is radial pulse is always absent in Takayasu's Arteritis? Does not one also get diminish pulsation also in thromboangiitis obliterans or diabetic arteriosclerotic obliterans? To answer just these few questions is enough to forget the name "Pulseless disease".

"Reverse Coarctation" is also commonly in usage. This is anatomically wrong. Coarctation denotes narrowing of the lumen of an artery, in general sense, of the aortic arch resulting in diminishing pulse in the lower extremities and a full radial pulse. In majority, the narrowing zone of the aorta is located distal to the insertion of the left subclavian artery. How can, anatomically, the coarctation be reverse in Takayasu's Arteritis? Where is the reversed part located in the aorta?

"Young Female Arteritis," is uninformative. Nothing can be obtained from this name except that there is an inflammation of an artery in a young woman. If this name continue to exist, we will have also a young man arteritis, old man arteritis, infantile arteritis, and so on.

“Mid-Aortic Syndrome” is more vague than Aortic Arch syndrome. It is difficult to form an idea where really the mid portion of the aorta is and what type of lesion involving that particular part of the aorta that produces the syndrome. Therefore, from anatomical point of view, this term is uninformative and unscientific.

“Marterell’s Syndrome” is used by some Spanish speaking group of workers is indeed easily comprehensible. Actually what Marterell described had already been previously described under arteriosclerotic obliterations of brachiocephalic arteries.

There are several other titles which are equally unnecessary because the good and original one—the Takayasu’s Arteritis—is already there. What and Why is the objection to the title is beyond understanding. We have accepted many names in calling disease entities originally or primarily described by respective workers in medical literature and there is no reason why not to accept this term as such. What one would get from applying so many names to describe a single disease entity is really a big question. No wonder, medical literature nowadays, is filled of so many mystery.

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## เส้นเลือดแดงอักเสบ ของทากายาสึ

กับ

เอออร์ติค อาร์ช ซินโดรม

บทความเรื่องนี้ได้อภิปรายถึงการใช้นิยามต่างๆ ในการบรรยายโรคเส้นโลหิตแดงอักเสบ ซึ่งทากายาสึได้รายงานไว้ในปี 1908 จากการทบทวนรายงานต่างๆ ของโรคนี้หลายราย จากวารสารต่างประเทศ ผู้เขียนแนะนำให้เลิกใช้คำ เอออร์ติค อาร์ช ซินโดรมหรือชื่ออื่นๆ เสีย และใช้ชื่อ ทากายาสึ แทนทุกรายในกรณีที่โรคมีลักษณะทางคลินิก และทางพยาธิสภาพตรงกันที่ได้อธิบายไว้

### References

1. Takayasu, M., A case with peculiar changes of the central retinal vessels. Acta Soc. Ophth. Jap., 12: 554-556; 1908 (quoted English Translation by Judge and Associates, 1961)
2. Judge, R.D., Currier, R.D., Gracie, W.A. and Figley, M.M., Takayasu's Arteritis and the Aortic Arch Syndrome., Am. J Med., 32: 379-392; 1962
3. Shimizu, K and Sano, K.. Pulseless disease., J. Neuropath. Clin. Neurol., 1: 37; 1951
4. Harrison, C. V., Takayasu's Arteritis., Recent Advances in Pathology, 1960, 7th edition, J.A. Churchill, London, pp. 177-180
5. Caccamise, W.C. and Whitman, J.F., Pulseless disease, preliminary case report. Am. H.J., 44:629-633, 1952
6. Hirsch, M.S., Aikat, B.V. nad Basu, A.K., Takayasu's arteritis. Report of five cases with Immunologic studies. Bull. Johns Hopk. Hosp.. 115:29; 1964
7. Ask-Upmark. E. and Fajers, C.M., Further observations on Takayasu's syndrome Acta Med. Scand. 155:275-191; 1956
8. Wickbom, I., Arteriography in Brachiocephalic arteritis (Pulseless disease or Takayashu Syndrome). Acta Radiol. 48:321-329; 1957.



9. Ask-Upmark, E., On the pulseless disease out-side of Japan. *Acta Med. Scand.* 149:n60-178; 1954
  10. Koszewski, B.J. and Hubbard, T.F., Pulseless disease due to Branchial Arteritis, *Circulation*, 16:406-v10; 1957
  11. Mengis, C.L , Dubilier, W. and Barry, K.G., The Aortic Arch Syndrome of Takayasu. *Am H.J.*, 55: 435-442; 1958
  12. Ross, R.S. and Mc Kusick, V.A., Aortic Arch Syndromes; diminished or absent pulses in arteries arising from the arch of the aortic. *AMA, Arch Int. Med.*, 92:701-740; 1953
  13. Thurlbeck, W.M. and Currens, J.H., The Aortic Arch Syndrome (pulseless disease): a report of ten cases with 3 autopsies. *Circulation*, 19:499-510; 1959
  14. Paloheimo, J.A., Julkunen, H., Siltanen, P. and Kajander, A., Takayasu's Arteritis and Ankylosing spondylitis *Acta Med. Scand.*, 179: 77-85; 1966
  15. Sandring, H and Welin,G., Aortic Arch Syndrome with special reference to Rheumatoid Arthritis. *Acta Med. Scand.*, 170:1-14; 1961
  16. Segal, F. and Berezowski, A., Aortic Arch Syndrome. *Am. H.J.*, 55:443-449; 1958
  17. Martorell, F. and Fabre, J., The syndrome of obliteration of the supra-aortic branches. *Angiology*, 5: 39-42; 1964
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