

# Cystic medianecrosis

Cystic medianecrosis (also called Erdheim's disease, cystic medial necrosis or cystic medial degeneration) is a disease affecting the large arteries, especially the aorta, which is characterized by **accumulation of mucopolysaccharides in the media of these vessels and** 'loss of muscle and elastic fibers. *These lesions resemble cysts in appearance.*<sup>[1]</sup>

In most cases, cystic medianecrosis occurs independently, but not infrequently also within congenital diseases of the connective tissue, e.g. Marfan's syndrome, Ehlers-Danlos syndrome or annuloaortic ectasia. The cause of this disease is unknown. An **autosomal dominant** inheritance is assumed.<sup>[2]</sup>

Despite its name, the lesions described in cystic medianecrosis are not always necrotic<sup>[3]</sup> and do not always form cysts<sup>[4]</sup>.

Cystic medianecrosis increases the risk of developing **aneurysm** or **aortic dissection**.

## Links

### External links

- Cystic medianecrosis (English Wikipedia) ([https://en.wikipedia.org/wiki/Familial\\_thoracic\\_aortic\\_aneurysm](https://en.wikipedia.org/wiki/Familial_thoracic_aortic_aneurysm))

### Related Articles

- Mucous dystrophy
- Intercellular mass disorders
- Mucopolysaccharidoses
- Inherited disorders of sugar metabolism

## References

1. Yuan, S. M., & Jing, H. (2011). Cystic medial necrosis: pathological findings and clinical implications. *Revista Brasileira de Cirurgia Cardiovascular*, 26(1), 107-115.
2. ERDHEIM CYSTIC MEDIAL NECROSIS OF AORTA, OMIM database. <https://www.omim.org/entry/607086>
3. Hirst, A. E., & Gore, I. (1976). Editorial: Is cystic medianecrosis the cause of dissecting aortic aneurysm?. *Circulation*, 53(6), 915-916.
4. Schlatmann, T. J., & Becker, A. E. (1977). Pathogenesis of dissecting aneurysm of aorta: comparative histopathologic study of significance of medial changes. *The American journal of cardiology*, 39(1), 21-26.



Microphotography showing a section through the wall of a vessel affected by cystic medianecrosis. The image shows numerous deposits of basophilic mass (upper part, blue) and disruption of elastic fibers. The tunica adventitia and vasa vasorum are seen in the lower part of the image.