

# Cystic medial necrosis

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

Cystic medionecrosis occurs in most cases independently, but often also within congenital connective tissue diseases, such as Marfan's syndrome, Ehlers-Danlos syndrome or anuloaortic ectasia. The cause of this disease is unknown. *Autosomal dominant* inheritance is assumed.<sup>[2]</sup>

Despite their name, the lesions described in cystic medial necrosis are not always necrotic<sup>[3]</sup> a netvoří vždy cysty<sup>[4]</sup>.

Cystical medial necrosis increases the risk of forming an **aneurysm** or an **aortic dissection**.

## Links

### External links

- Cystická medionekróza (anglická wikipedie) ([https://en.wikipedia.org/wiki/Familial\\_thoracic\\_aortic\\_aneurysm](https://en.wikipedia.org/wiki/Familial_thoracic_aortic_aneurysm))

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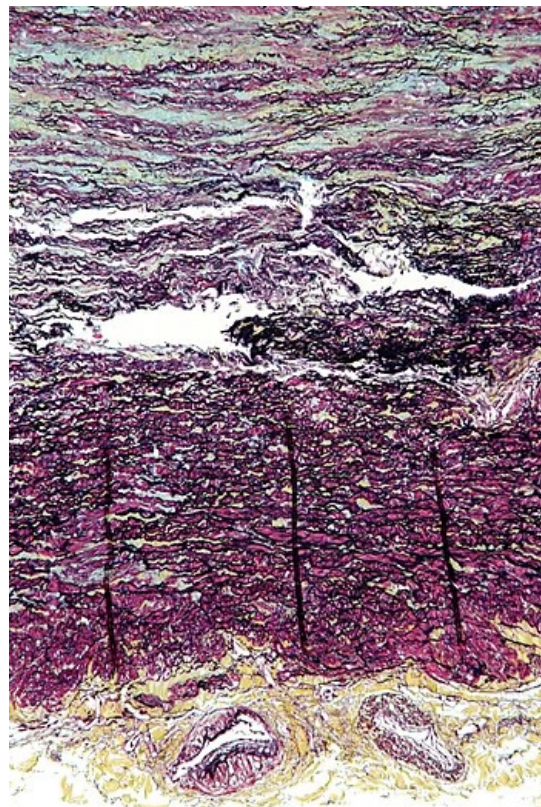
- Mucus dystrophy
- Disorders of intercellular mass
- Mukopolysacharidosis
- Hereditary disorders of carbohydrate metabolism

## 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, databáze OMIM. <https://www.omim.org/entry/607086>
3. Hirst, A. E., & Gore, I. (1976). Editorial: Is cystic medionecrosis the cause of dissecting aortic aneurysm?. *Circulation*, 53(6), 915-916.
4. Schlattmann, 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.

## Source

- [ws:https://www.wikiskripta.eu/index.php?curid=68571](https://www.wikiskripta.eu/index.php?curid=68571)



Microphotograph showing a section of a vessel wall affected by cystic medionecrosis. The picture shows numerous deposits of basophilic mass (in the upper part, blue) and failure of elastic fibers. The lower part of the image shows the tunica adventitia and the vasa vasorum.