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## CLINICOPATHOLOGIC CORRELATIONS

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### Pseudocoarctation, Kinking or Buckling of the Aorta

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#### SUMMARY

Pseudocoarctation, buckling, and kinking of the aorta are terms used to describe a roentgenologic deformity of the aorta similar to that in classical coarctation but in which no obstruction is demonstrable. It is probable that the first three terms used are synonyms for subclinical coarctation. In this state, the lesion of classical coarctation of the aorta is present and the two show the same associated anomalies.

THE TERM pseudocoarctation has varying connotations. To Dotter and Steinberg,<sup>1</sup> who introduced the term in 1952, it was intended to signify the same lesion as in classical coarctation yielding the same radiologic picture but with little or no obstruction. Others have implied that the state was different fundamentally from coarctation, representing a long aorta which was "kinked" or "buckled."<sup>2</sup>

In order to determine whether there is a fundamental difference between coarctation with mild or no obstruction and kinked or buckled aorta, it is proper first to define the lesion of classical coarctation and then to determine whether or not this lesion is present in cases which may be called kinked aorta.

Over a century ago, in 1841, Craigie<sup>3</sup> described several cases with varying degrees of localized obstruction of the aorta at the junction of the arch and its descending portions. Regarding the gross appearance of

the aorta at the involved site, his description of one of the cases is particularly significant. He stated, "At the ductus arteriosus the aorta exhibited a constriction similar to that which would occur if a sharp body had been pressed upon its upper surface until it had diminished the caliber of the lumen by one-half."

In 1948, one of us (J.E.E.) and others<sup>4</sup> reported on the anatomic features of classic coarctation. The basic lesion is represented by a localized ridgelike thickening of the aortic wall, usually lying at the junction of the arch and descending aorta. The ridge involves the superior, posterior, and anterior aspects of the aorta, while the lower wall, that into which the ductus arteriosus or ligamentum arteriosum inserts, is not involved. Grossly, the ridge projects into the lumen causing it to be narrow, eccentric, and to lie near the lower wall of the aorta (fig. 1a). The outer wall of the aorta shows a sharp concavity along the superior wall which corresponds with the position of the ridge and the site at which "a sharp body had been pressed," according to Craigie (fig. 1b). Histologically, the ridge is composed of medial tissue (fig. 2).

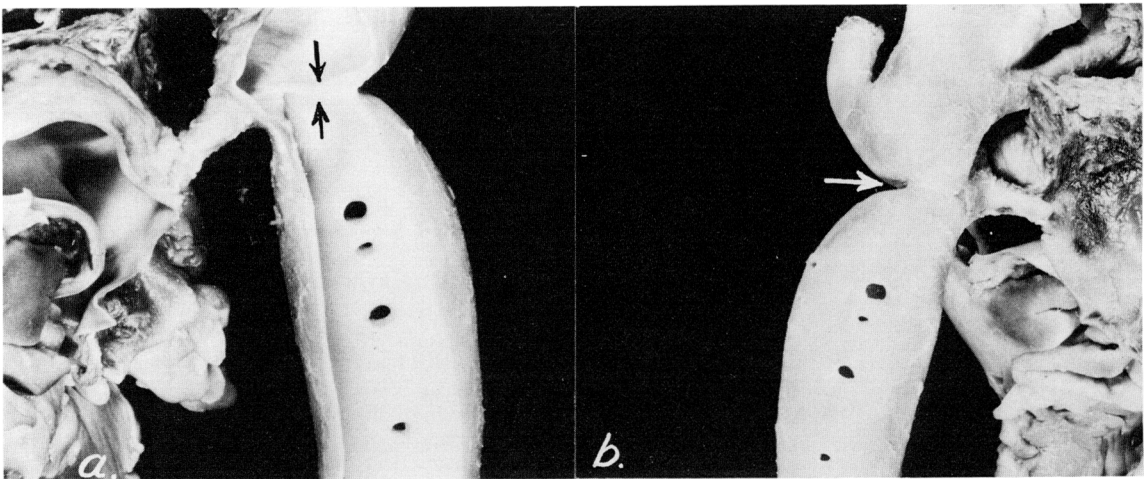
Although the term coarctation was originally intended to indicate constriction of the aorta, it now may be viewed in a broader sense; that is, that the lesion of coarctation has certain features and in its presence the degree of aortic obstruction may vary.

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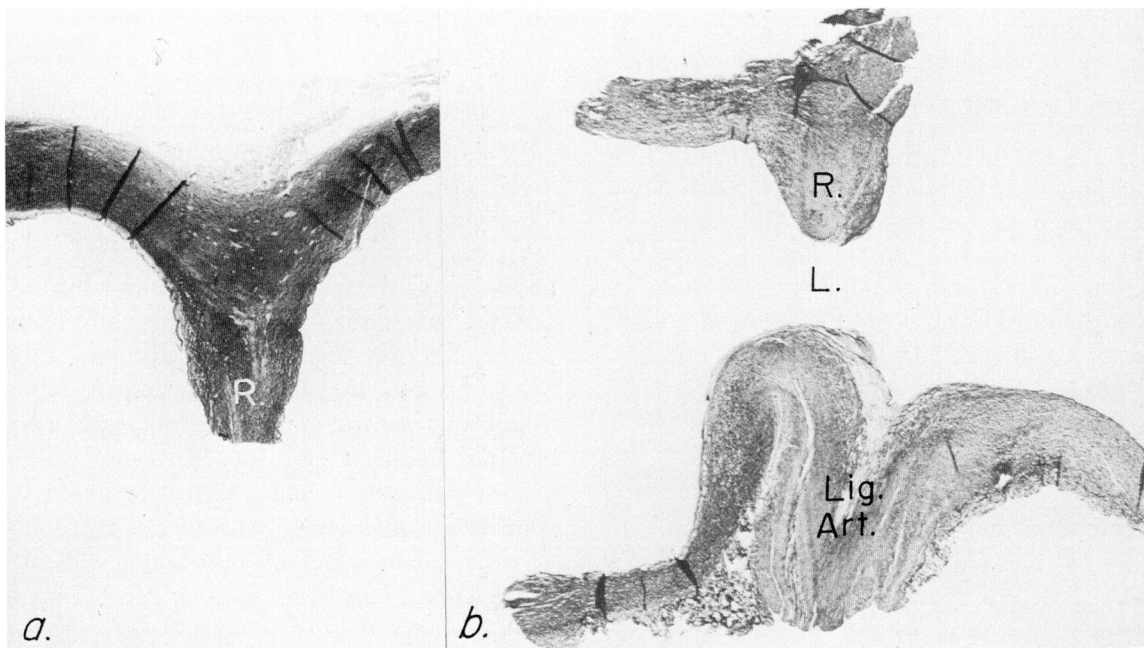
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**Figure 1**

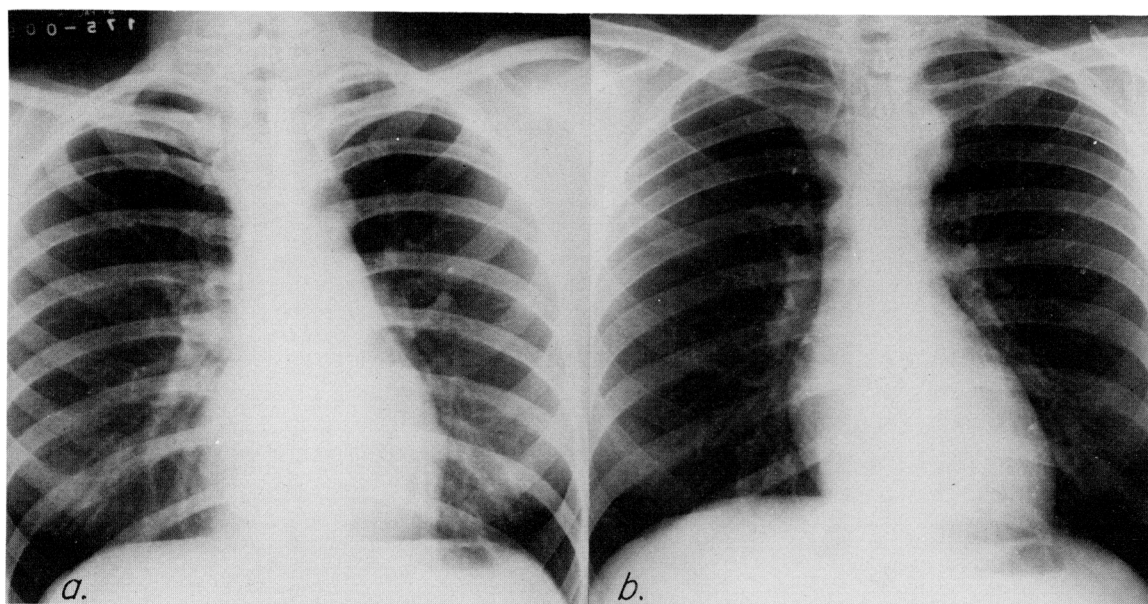
*Classical coarctation of the aorta. (a) Interior of aorta viewed from front. Corresponding with the indentation in the superior wall is a ridge (between arrows) which protrudes into the lumen causing the latter to be narrowed. (b) Aorta viewed from behind. At the junction of the arch and descending portions is a characteristic sharp indentation (point of arrow) in the superior aspect of the vessel.*



**Figure 2**

*Photomicrographs in classical coarctation of aorta. (a) Superior wall of aorta from the case shown in figure 1. The ridgelike protrusion (R.) is composed of medial tissue which projects into the lumen. Elastic tissue stain  $\times 6.5$ . (b) Surgically resected segment of coarctation of aorta viewed from front. Slightly proximal to the junction of the ligamentum arteriosum (Lig. Art.) with the aorta is a ridge (R.) involving the superior aspect of the vessel. The latter protrudes into the lumen (L.) and causes it to be narrow and eccentric. Elastic tissue stain  $\times 8$ .*





**Figure 3**

*Roentgenograms each showing the "3 sign" of coarctation. (a) From a case of classical coarctation. Notching of the ribs is an associated finding. (b) From a case in which no demonstrable obstruction of the aorta was evident, the case being an example of kinked aorta or pseudocoarctation. Notching of ribs is absent.*

The indentation of the superior aspect of the aorta, coupled with wide segments proximally and distally, yields the roentgenographic picture of the "3 sign" seen in classic coarctation (fig. 3a). The aortic deformity may suggest a thoracic tumor rather than a vascular anomaly.<sup>5</sup> Notching of the ribs, which may be a companion sign of classical coarctation, represents the effects of collateral circulation incident to significant aortic obstruction.

There are cases with the same roentgenographic appearance of the aorta as in classical coarctation but which show neither measurable aortic obstruction nor notching of the ribs. Such cases bring up the diagnosis of pseudocoarctation or buckling of the aorta (figs. 3b and 4) as well as thoracic tumor.

Since the hazards of aortic obstruction are absent, pathologic material from such cases is scanty; nevertheless, some is available.

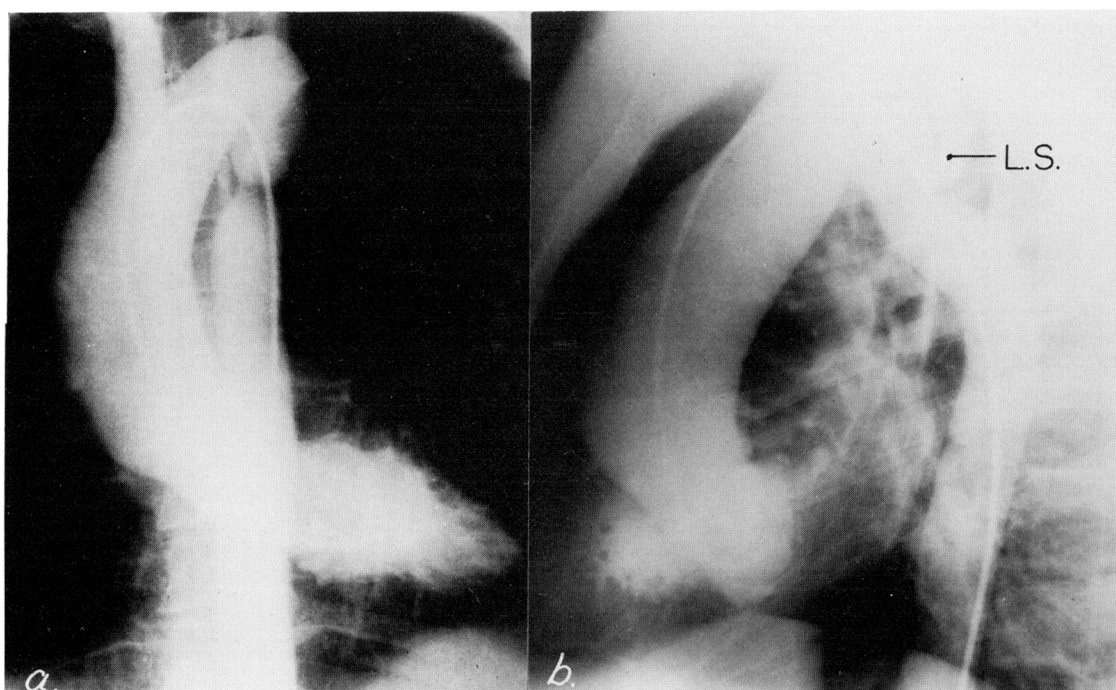
In 1962, Hagstrom and Steinberg<sup>6</sup> indicated that pseudocoarctation is a subclinical form of coarctation. The fact that the lesion of

coarctation is variable in the degree of narrowing it causes is supported by the observations of Hamilton and Abbott.<sup>7, 8</sup> In 1928, these authors graded coarctation in three categories: atresia; extreme, lumen less than 5 mm; and moderate, lumen greater than 5 mm.

In our experience, cases with the clinical definition of pseudocoarctation or kinked aorta qualitatively show the aortic lesion of coarctation (fig. 5). At the junction of the arch and ascending aorta, the external aspect of the superior aspect of the vessel shows a localized convexity.

Internally and corresponding with the position of the indentation involving the superior aspect is a shallow ridge (fig. 5c). The ridge becomes more shallow as it arches onto the anterior and posterior walls of the aorta. It does not involve the inferior wall. The effect of the shallow ridge upon the aortic channel is slight.

Histologic examination of the aortic wall at the level of the ridge shows a focus of medial



**Figure 4**

*Aortograms in pseudocoarctation or kinked aorta. (a) Frontal view. (b) Lateral view. An indentation in the superior wall of the aorta is present just distal to the origin of the left subclavian artery (L.S.). The proximal aorta is tortuous, as is the aorta distal to the indentation. Pathologic specimen shown in figure 5. Bicuspid aortic valve was associated in this case.*

thickening qualitatively like that in classic coarctation of the aorta (fig. 5d). It may be the case that poststenotic dilatation of the aorta is an additive factor in yielding the classic roentgenographic shadow.

Accepting that there is a variant of coarctation that may be termed pseudocoarctation, Edmunds and associates<sup>9</sup> stated that pseudocoarctation must be distinguished from kinking or buckling. They considered kinking or buckling of the aorta different from pseudocoarctation but did not define the difference. It is possible that simple elongation of the aorta may yield a picture of buckling which, in turn, may be confused with pseudocoarctation. While we cannot deny this categorically, we have not been able to confirm such a claim.

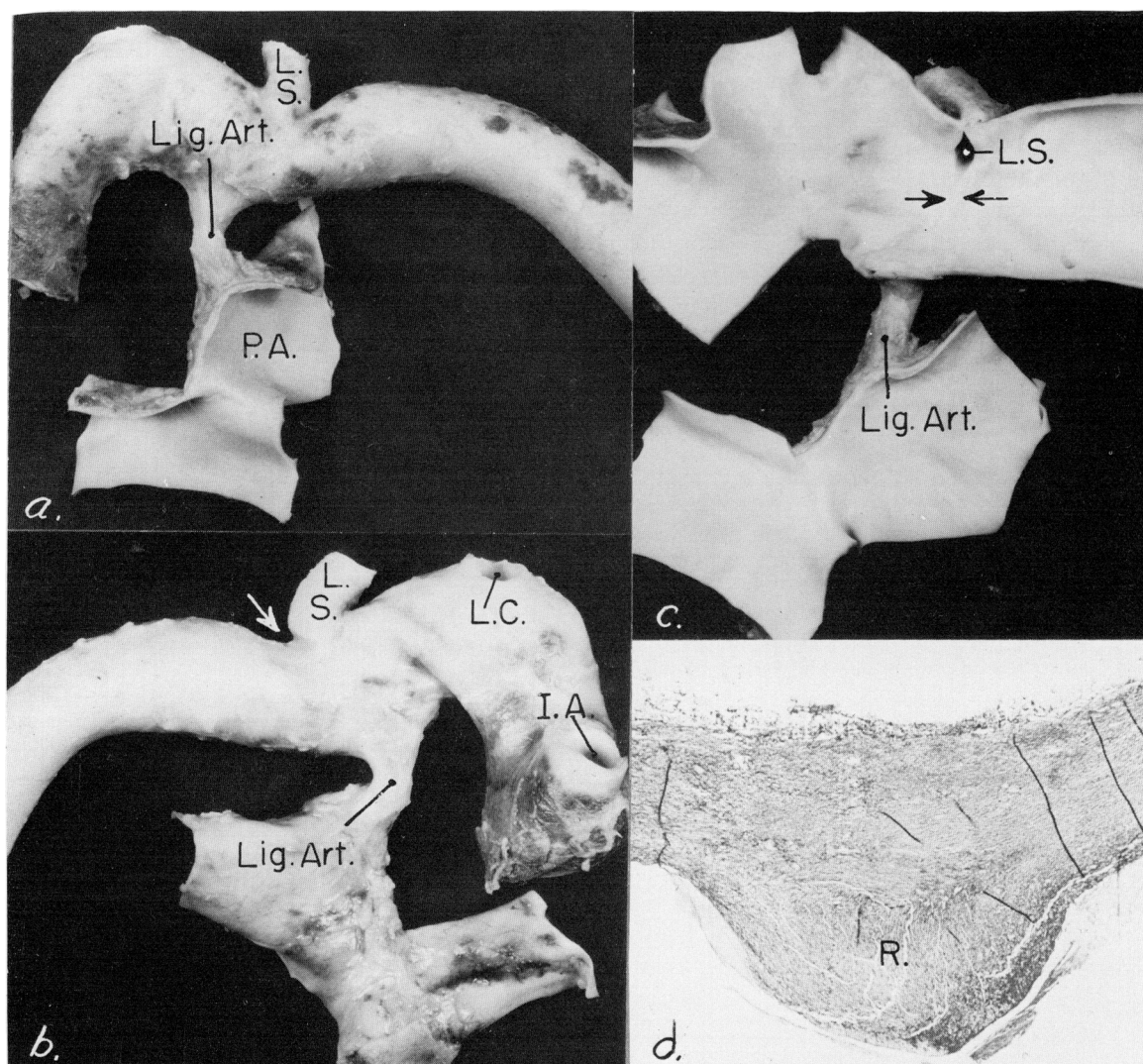
It is therefore our opinion that pseudocoarctation, on one hand, and so-called kinking or buckling of the aorta, on the other, are terms for the same lesion. To our knowledge, there

has not as yet been presented a pathologic description in cases with the roentgenographic picture under discussion in which the basic lesion is not that of coarctation. Until such a condition is met, we believe that cases designated as pseudocoarctation are not different from those chosen to be called kinking or buckling.

Supporting evidence for the concept that pseudocoarctation or buckling is a minor degree of classical coarctation comes from the fact that the two conditions share the same associated anomalies such as congenital bicuspid valve, patent ductus arteriosus, and subaortic stenosis.<sup>10-12</sup>

With the foregoing in mind, it seems appropriate to make the following generalizations. The aorta may be the site of a medial deformity which may be called the lesion of coarctation. From case to case, this lesion may cause either no obstruction or degrees of





**Figure 5**

Pathologic specimen from case of pseudocoarctation of which the aortogram is shown in figure 4. (a) The thoracic aorta viewed from front. Immediately distal to the origin of the left subclavian artery (L.S.) is an indentation in the superior aspect of the aortic wall. This lies distal to the ligamentum arteriosum (Lig. Art.); the latter is shown arising from the pulmonary artery system (P.A.). (b) Aorta, pulmonary arterial bifurcation, and ligamentum arteriosum (Lig. Art.) viewed from behind. The left subclavian artery (L.S.) lies just proximal to an indentation (arrow) in the superior aspect of the aorta. The left common carotid (L.C.) and innominate (I.A.) arteries are relatively distant from the left subclavian artery and from each other, suggesting an increased length to the aorta. (c) Interior of aorta viewed from front. Just distal to the insertion of the ligamentum arteriosum (Lig. Art.) and immediately distal to the origin of the left subclavian artery (L.S.) the superior aspect of the aorta shows a ridge (between arrows) which becomes less evident as it arches toward the insertion of the ligamentum arteriosum. (d) Photomicrograph of the superior wall of the aorta at level between arrows. Characteristic deformity of the aortic wall in which a ridge (R.) is formed by medial tissue. The picture is similar to that in classic coarctation as shown in figure 2. Elastic tissue stain  $\times 12$ .

obstruction varying from mild to severe. Although terms like pseudocoarctation, kinking, or buckling are convenient, one might consider that there is a range of degrees of obstruction among subjects with the basic lesion. One could therefore classify cases of coarctation as showing no, mild, moderate, or severe degrees of aortic obstruction. Otherwise, confusion will reign in the area of mild degrees of coarctation with respect to its pathology and associated anomalies.

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