

Embryological Significance of Lymphangioma*

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Since 1938 when Goetsch studied injected surgical specimens, the growth of cystic hygromas has always been considered as a continuous budding from the walls of the tumour cavity.

This phenomenon was compared to the foetal centrifugal development of peripheral lymphatics from the walls of the primary lymphatic sacs, in keeping with Florence Sabin's theory of lymphatic embryology (1912).

Although the microscopical observations made by Goetsch were remarkable, their interpretation ought to be revised, as Sabin's theory has since proved wrong. Her observations on embryos were made by injecting primary lymphatic sacs with a dye. The central lymphatic system was filled, showing several large cavities. At a later stage more peripheral networks were filled, and these were interpreted as ramifications from the walls of the primary lymphatic sacs (Fig. 1). This theory must be discarded, as it was based upon retrograde injection, a method that is not adequate for studying the development of a vascular system.

Lymphatic vessels do not generate by centrifugal growth from veins and lymph cavities. At a very early stage (9 to 12 mm.) of foetal development, mesenchymal slits appear in the reticulum of a rich venous plexus (Fig. 2A). By coalescence of these spaces, large lymphatic cavities are formed (Fig. 2B) which open secondarily into the venous system (Fig. 2C). Later, they decrease in size and straighten out, following the direction of the veins (Fig. 2D).

This theory of the lymphatic system development was put forward by Huntington (1911) and confirmed for humans by Kampmeier (1931), who studied serial sections and reconstructions of human embryos (Fig. 2).

Should the primary lymphatic spaces fail to join the central system, cystic formation is induced. The lack of communication between a lymphatic sac and the venous system produces a cystic hygroma. This is the reason why cystic hygromas are found in

the same locations as foetal lymphatic sacs—cervical, mediastinal, or retroperitoneal.

In cavernous lymphangioma the sequestration is more peripheral than in cystic hygroma: primary lymphatic spaces do not join the main central collectors. Lymphangioma simplex is the result of a still more localized sequestration of a few mesenchymal slits.

Cystic hygroma and cavernous lymphangioma are thus essentially the same: both are multilocular

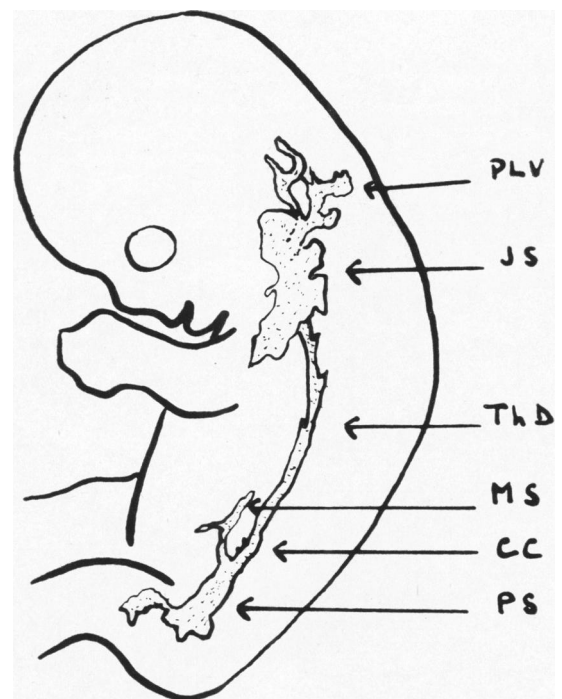


FIG. 1.—Human embryo of 30 mm. JS, jugular sac; ThD, thoracic duct; CC, cysterna chyli; MS, mesenteric sac; PS, posterior sac; PLV, peripheral lymphatic vessels. (After Sabin, 1912.)

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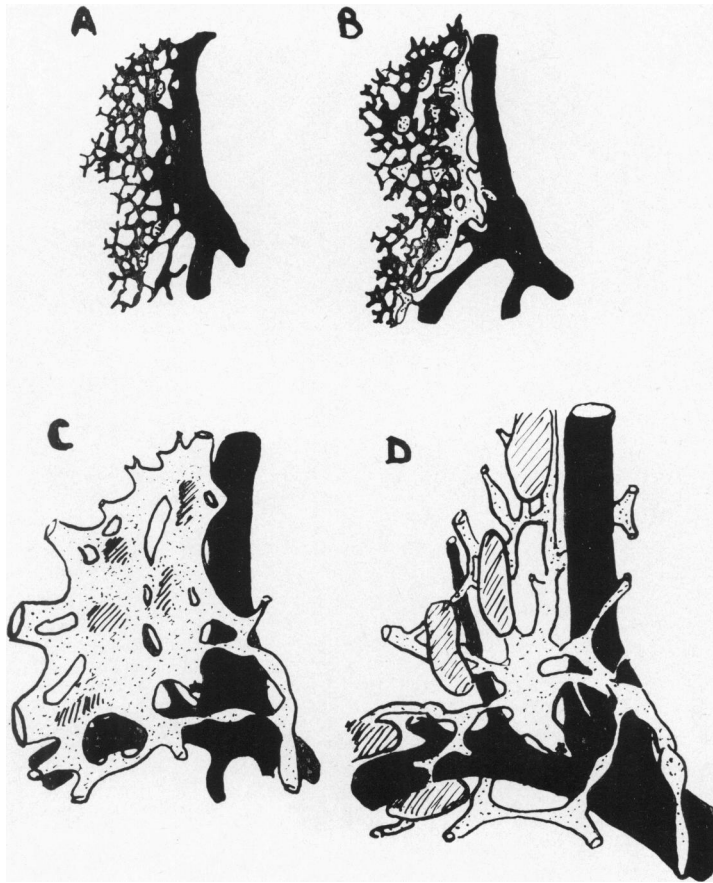


FIG. 2.—Human embryo: cervical region. Successive stages in development of lymphatic system (see text).

- (A) Venous plexus; a few mesenchymal slits (dotted spaces).
 (B) Coalescence of the lymphatic spaces.
 (C) Jugular lymphatic sac opening into the venous system.
 (D) Narrowing of the lymphatic vessels; lymph nodes. (After Kampmeier, 1931.)

cavities filled with lymph and lined with endothelium (Bill and Sumner, 1965). In the former, these cavities are large; in the latter they are smaller and numerous (Fig. 3, *overleaf*). An association of both forms may be seen with gradual transition of one form into the other.

The tumour grows because the atresia of the main collectors prevents its emptying. The volume of the tumour usually increases after inflammation because the fluid balance between arteries and veins is disturbed and filtration and lymph formation are consequently increased. The accumulated fluid separates bundles of muscle fibres, vessels, and nerves, which are soon surrounded by fluid and which degenerate under fluid pressure.

Conclusion

The pathogenesis of lymphangiomas must be considered in the light of current concepts of lymphatic development. Tumour growth occurs through accumulation of peripheral lymph trapped in the central cavities, secondary to atresia of the main collectors (Fig. 3, *overleaf*).

REFERENCES

- Bill, A. H., Jr., and Sumner, D. S. (1965). A unified concept of lymphangioma and cystic hygroma. *Surg. Gynec. Obstet.*, **120**, 79.
 Godart, S., Collette, J., and Dalem, J. (1964). Pathologie chirurgicale des vaisseaux lymphatiques. *Acta chir. belg.*, Suppl. 1, 5.



FIG. 3.—Lymphangiogram of a congenital lymphoedema or lymphangioma of the left leg. Lipiodol adheres to the walls of lymph cavities. No collector is present on the left side. Normal lymphangiogram on the right side. (Godart, Collette, and Dalem, 1964.)

Goetsch, E. (1938). Hygroma colli cysticum and hygroma axillare. Pathologic and clinical study and report of 12 cases. *Arch. Surg.*, **36**, 394.

Huntington, G. S. (1911). *The Anatomy and Development of the Systemic Lymphatic Vessels in the Domestic Cat*. Wistar Inst. Anat. & Biol., Philadelphia.

Kampmeier, O. F. (1931). Ursprung und Entwicklungsgeschichte des Ductus thoracicus nebst Saccus lymphaticus jugularis und Cisterna chyli beim Menschen. *Morph. Jb.*, **67**, 157.

Sabin, F. R. (1912). The development of the lymphatic system. In *Manual of Human Embryology*, ed. F. Keibel and F. P. Mall, Vol. 2, p. 709. Lippincott, Philadelphia.