ROLE OF SPLENECTOMY IN DIAGNOSIS AND MANAGMENT IN PEDIATRIC PRACTICE IN HEMATOLOGICAL DISORDERS

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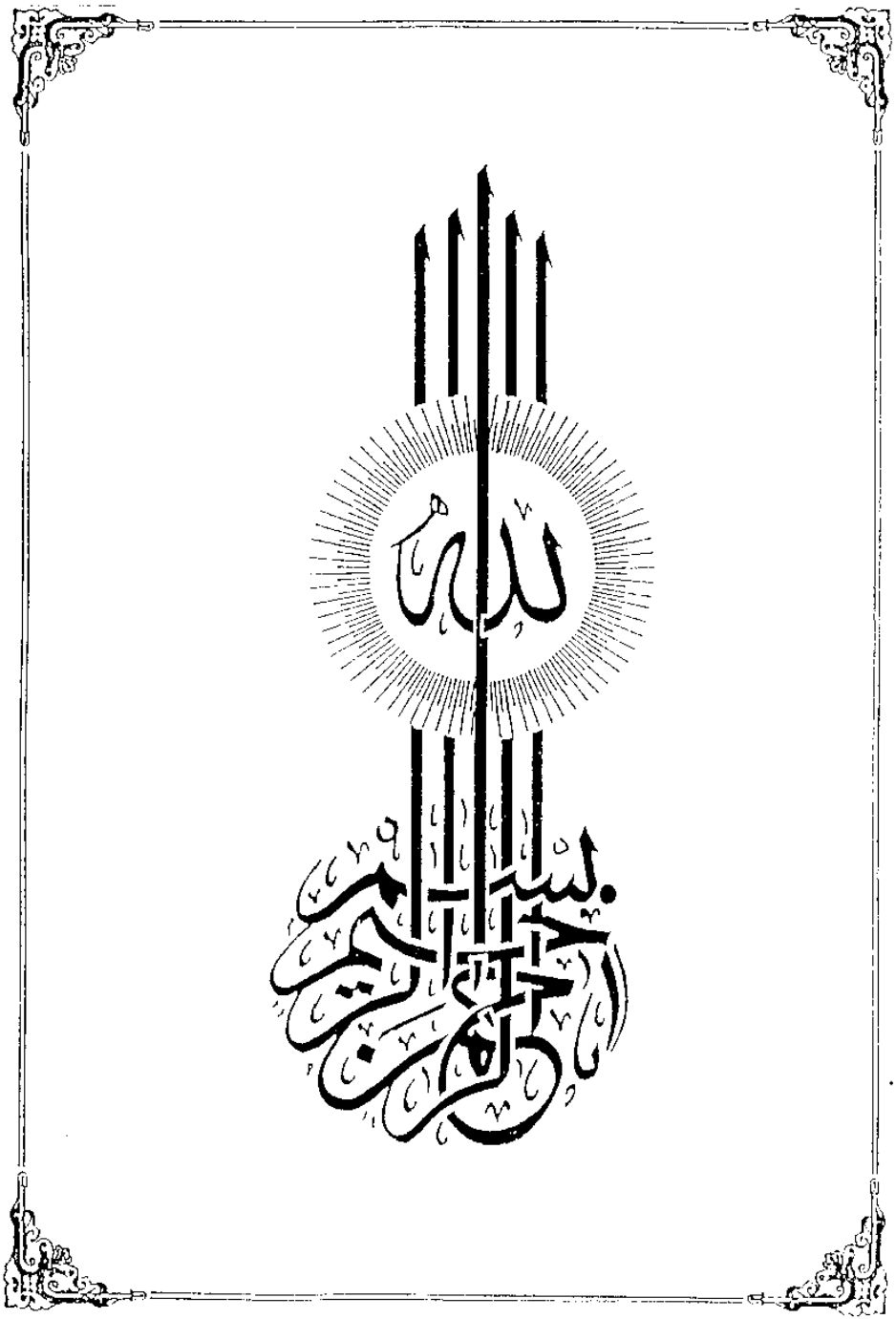

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INTRODUCTION

Splenomegaly is encountered among Egyptian children due to various etiology. Among causes of enlarged spleen are; parasitic infestation (Schistosomiasis, Malariasis), blood diseases (Beta Thalassemia, Hemoglobinopathies, Idiopathic Thrombocytopenic purpura (I.T.P.), Hereditary Spherocytosis), and Lymphomas (Whether Hodgkin's or non Hodgkin's).

Examination of spleen by histological and pathological sections may be required for diagnosis of cases and for deciding about treatment (e.g. staging of Hodgkin's disease), on the other hand, splenectomy might be required as a mode of treatment; to prolong periods between subsequent transfusions in Thalassemia Major or to cure I.T.P. and Hereditary Spherocytosis. Also splenectomy is indicated in cases of hypersplenisim and as a step of operations for portal hypertension.

AIM OF THE WORK

This work is concerned to study the role of splenectomy in diagnosis and managment in pediatric practice in cases with hematological disorders.

EMBRYOLOGY AND ANATOMY

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The spleen appears about the sixth week as a localised thickening of the coelomic epithelium of the dorsal mesogastrium in its cephalic portion, and the proliferating cells invade the underlying mesenchyme, which has become condensed and vascularised. The process occurs simultaneously in several adjoining areas which soon fuse to form a lobulated spleen, derived in part from the coelomic epithelium and in part from the mesenchyme in the dorsal mesogastrium. The earlier lobulated character of the organ disappears but is indicated by the presence of notches on its upper border in the adult.

The spleen is situated principally in the left hypochondriac region of the abdomen. It varies in shape, according to the size of the colic impression, from a slightly curved wedge to a tetrahedron. In the adult it measured 1 x 3 x 5 inches (nearly as the closed fist of the hand), weights between 75-100gm, decreasing somewhat with age. Its long axis lies in the line of the tenth rib, its posterior extremity being about 3.5 to 4cm. from the mid-dorsal line opposite the spine of the tenth vertebra, and its anoterior extremity reaches the mid-

axillary line. It is soft, of friable consistence, highly vascular, and of dark purplish colour.

The spleen has a convex, smooth, diaphragmatic surface. It is in relation with the abdominal surface of the diaphragm, which separates it from the lowest parts of the left lung and pleura and the ninth, tenth and eleventh ribs of the left side.

Also, it has a visceral surface which is directed towards the obdominal cavity, and presents gastric, renal, pancreatic and colic impressions.

The gastric impression is broad and concave. It is in contact with the posterior wall of the stomach, from which it is separated by a recess of the greater sac of the peritoneum. It presents near its lower limit the hilus which is pierced by several irregular apertures for the entrance and exit of vessels and nerves.

The renal impression, which is slightly concave, is related to the upper and lateral part of the anterior surface of the left kidney and, sometimes, to the upper pole of the left suprarenal gland.

The colic impression, which is usually flattened, is related to the left colic flexure and phrenicocolic ligament.

The pancreatic impression, when present, is small and is directly related to the tail of the pancreas which lies in the lienorenal ligament.

The spleen has two borders, the superior border which separates the diaphragmatic surface from the gastric impression. It is marked, near its lateral end, by one or two notches of variable depth. The inferior border separates the renal impression from the diaphragmatic surface. It corresponds to the lower margin of the eleventh rib.

The posterior extremity is directed towards the vertebral column. The anterior extremity is related to the left colic flexure and the phrenicocolic ligament.

The spleen is almost entirely surrounded by peritoneum, which is firmly adherent to its capsule. Recesses of the greater sac intervene between the spleen and the stomach, and between the spleen and the left kidney. It develops in the upper part of the dorsal mesogastrium and remains connected with the stomach and the posterior abdominal wall by two folds of peritoneum - One, termed the lienorenal ligament, the splenic vessels pass between its two layers. The other fold, termed the gastrosplenic ligament, also consists of two layers, the short gastric

and left gastro-epiploic branches of the splenic artery run between its two layers.The lateral part of the lateral end of the spleen is in contact with the phrenicocolic ligament.(Gray's Anatomy, 1976).

Accessory spleens have been reported in 14-30 % of patients with a higher incidence occuring in patients operated on for hematologic disorders. These accessory organs, which receive their vascular supply from the splenic artery, are present, in decreasing order of frequency, in the hilus of the spleen, the gastro-splenic and splenocolic ligaments, the gastrocolic ligament, the splenorenal ligament, and the greater omentum. They also may occure in the pelvis of the female, and functioning splenic tissue has been removed from the scrotum in juxtaposition to the left testicle (Olsen and Beaudoin. 1969).

The blood supply of the spleen is provided by the splenic artery. It passes between the layers of the lieno-renal ligament and at the hilum of the spleen breaks up into a four or five branches which enter the hilum separately. Similar veins leave the hilum and unite to form the splenic vein, The lymph drains into several nodes lying at the hilum and thence by way of the retropancreatic nodes, to the coeliac lymph nodes. The nerves are derived from the coeliac plexus and distributed mainly to the

branches of the splenic arteries.

The spleen is enclosed in a dense fibromuscular capsule 1-2mm thick, from which trabeculae penetrate the substance of the organ and enclose the pulp. A bloody pulp fills the cavities between the trabeculae; there is no subcapsular space.

The capsule and trabeculae consist of collagenous white fibrous tissue and yellow elastic fibres, the latter being more numerous in the trabeculae. The pulp consists of all varities of white cells, small and large, monocytes, histiocytes, and multinucleated giant cells, lying on and between a network of reticulin fibres; this is an important part of the reticuloendothelial system. Free erythrocytes abound, mixed with the conglomeration of white cells.

A most characteristic feature of the spleen is the presence of Malphighian corpuscles. These consist of aggregates of lymphocytes just visible to the naked eye. Each is perforated by a small arteriole, though the arteriole is not seen in every Malphighian follicle cut in histological section, for the following reason. The branches of the splenic artery run in the trabeculae and then branch into the splenic pulp. Each breaks up into a leash of straight vessels that enter blood sinsoids in the pulp;

Each straight vessel is invested with a spherical mass of lymphocytes; the spleen is thus studded with masses of separate lymphatic follicles, each perforated by a straight arteriole. A section across the arteriole will show it lying within the lymphatic follicle; a section parallel with the arteriole will show the lymphatic follicle and no arteriole. The straight arterioles run in all directions, and on the law of averages it is necessary to inspect at most half a dozen follicles to find one showing the cut central arteriole. This arrangement is unique to the spleen; nowhere else in the body does an arteriole lie within a lymphatic follicle.

The straight arterioles open into thin-walled sinusoids which in turn drain into venules. There is no anastomosis between adjacent arterioles, i.e., the arteries themselves are end arteries and their obstruction leads to infarction of the spleen.

Small arteriolar branches of the splenic arteries leave the trabeculae, and their adventitial coat becomes replaced by a sheath of lymphatic tissue which accompanies the vessels and their branches almost as far as their division into capillaries. These lymphatic sheaths constitute the white pulp of the spleen, and here and there the sheaths are enlarged to form the splenic lymphatic follicles (Malphighian bodies), which vary in diameter

from 0.25mm. to 1mm and are visible to the naked eye on the freshly cut surface of the organ as whitish semiopaque dots contrasting with the dark red colour of the surrounding tissue, which constitutes the red pulp of the spleen. These lymphoid follicles are centres of lymphocyte production. There are no lymph vessels in the follicles and the lymphocytes migrate by amoeboid movement into the surrounding red pulp and continue to multipl there. The follicles atrophy with increasing age and may be absent in old age. The arterioles usually occupy an eccentric position in the follicles and give off fine side branches to supply the follicles. The arterioles finally lose their sheaths of lymphatic tissue and enter the surrounding red pulp, where they at once divide into a number of straight vessels termed penicilli. Each of these straight vessels, after running a course of about 0.5mm., shows a slight thickening of its coat, known as an ellipsoid, which is formed by a condensation of reticular cells and macrophages, and the lumen of the vessel is considerably narrowed. Beyond the ellipsoids, each vessel continues as an arterial capillary, or may divide into two capillaries, the termination of which is undecided.

The spleen is the most vascularised organ in the body, and the splenic circulation has been the subject of disagreement for almost 300 years. Three major concepts of splenic circulation

have been proposed. The "closed circulation" theory holds that, the capillaries are directly contineous with venous sinusoids that lie in the red pulp, and these in their turn convey the blood to the venous tributaries of the splenic veins that lie in the trabeculae and eventually emerge from the organ at the hilus, the endothelial coat of the capillaries, sinusoids and veins being continuous so that the blood does not come into direct contact with extra-vascular structures. This theory generally has been abandoned because of the proven nature of the sinus walls, which as early as 1686 were shown by Malphigi to contain perforations in the finest ramifications of the intrasplenic veins. The "open circulation" theory, which can be credited to Billroth in (1860), suggests that the arteries empty into tissue spaces or that sheathed capillaries pour their blood directly into pulp cords from which the blood can then pass through sinus wall stomata and enter the veins. The theory of "divided circulation", as proposed by Kinsely, suggests that whole blood enters the sinuses from the afferent capillaries and that deplasmatization occurs through the sinus walls. Some adopt a "compromise" theory and claim that, whereas the circulation may be 'open' in the distended spleen, it becomes functionally 'closed' in the contracted organ, while others maintain that the lining cells of the sinusoids are extremely sensitive and that even with a 'closed