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SPLENECTOMY IN PAEDIATRIC AGE GROUP

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Abstract

Background: In view of splenectomy in dealing with certain clinical problems in paediatric age group.

Objective: To describe the profile, indications, post-operative management and outcomes of children undergoing splenectomy.

Methods: All children undergoing splenectomy from June'2019 till August'2021 at INDEX MEDICAL COLLEGE, INDORE, M.P.

Results: The mean age at surgery was 9.9 years (range 3-16). Most splenectomies were per-formed for haematological disorders and were open .The mean post-operative length of stay (LOS) in patients who underwent open surgery was 4 days. No cases of overwhelming post splenectomy infec-tion (OPSI) were noted. At study completion, haematological disorders were the most common indication for splenectomy in children.

Conclusion: Indications for paediatric splenectomy mirror those found in international litera-ture. We haven't performed splenectomy, laparoscopically. Within this short span of time, no mortal-ity was recorded. We did not have any case of OPSI.

Key words: splenectomy

Introduction

Splenectomy is a well-described procedure in children. As opposed to the adult literature, trauma is an uncommon indication for splenectomy in children[1]. Frequently, failure of medical therapy alone to control the splenic sequelae of haematological disorders, e.g. splenomegaly, hypersplenism and massive infarction, necessitates splenectomy. Common examples of haematological disorders that necessitate splenectomy in children are Thalassemia, hereditary spherocytosis (HS), idiopathic thrombocytopae-nic purpur (ITP) and sickle cell disease (SCD). Although the pre-, peri- and postoperative manage-ment of paediatric patients undergoing elective and emergency splenectomy is well described within the international literature[2]. The objective of our study was to describe the profile, indications, post-operative management and outcomes of children undergoing splenectomy at INDEX MEDICAL COLLEGE HOSPITAL AND RESEARCH CENTRE, INDORE, M.P. between JUNE 2019 and AUGUST 2021.

Methods:

Study of paediatric patients undergoing splenectomy at INDEX MEDICAL COLLEGE HOSPITAL

RESEARCH CENTRE, INDORE, M.P. between JUNE 2019 and AUGUST 2021 was conducted. All patients between the ages of 0 and 16 years who underwent splenectomy were included in the study. Demographic data, data on indications for splenectomy, postoperative medical management, postoperative complications were recorded.

Results:

A total of 22 splenectomies were performed on children between JUNE 2019 and AUGUST 2021. 48% patients undergoing splenectomy were male, while 52% were female. The mean age at surgery was 9.9 years. The majority of the splenectomies were performed for haematological disorders like Thalessemia and the remainder were performed due to left sided portal hypertension, vascular malformation, splenic torsion, splenic abscess, trauma or with pancreatic tumour surgery. Table 1 describes the specific indications for surgery in our series.

The use of splenectomy in thalassemia has declined in recent years. Splenectomy is indicated in the trans-fusion-dependent patient when hypersplenism increases blood transfusion requirement. Splenectomy is a very effective approach for the management of selected children with primary ITP as evidenced by platelet count recovery. Although low, the risk of adverse events both peri-operative and in the long-term should be carefully considered when indicating this therapeutic modality.

Splenectomy in Herediatary Spherocytosis usually results in disappearance of anaemia and a clear de-crease of haemolytic markers . Patients with CDA underwent splenectomy also showed a moderate increase in hemoglobin concentration (9.3±1.2 g/dL to 10.6±1.6 g/dL).

Splenectomy is the treatment for recurrent acute splenic sequestration crises and hypersplenism in SCD, there is no evidence that it increases haemoglobin level, decreases haemolysis or improves pa-tients' survival. All patients in our series underwent total splenectomy.

LSPH should be considered in the presence of gastrointestinal bleeding with normal liver function tests and unexplained splenomegaly. Treatment should be directed to the underlying diseases, and while splenectomy is the treatment of choice for cases complicated by variceal bleeding.

In case of WS acute torsion, the decision making in children is difficult. The option to preserve the spleen would seem preferable to avoid the risk of sepsis, however it has to be balanced with the risk of prolonged thrombocytopenia, multiple transfusions and a possible second procedure to remove the spleen.

Haemangiomas are generally not treated unless they are symptomatic or very large, with an increased risk of haemorrhage; treatment then is usually a splenectomy.

Meticulous search for SPLENICULI was done and was removed at the time of surgery

The relevant complications are summarised in Table 2. All patients undergoing splenectomy received immunisations against Streptococcus pneumoniae, Haemophilus influenzae, and Meningococcus. Booster immunisations were given at follow-up where necessary.

ORAL PENTID was provided 2-3 weeks prior the planned splectomy at our setup.

Table 1: Indications for Splenectomy

ITP.	7 patients
THALESSEMIA	3 pateints
HEREDIATARY SPHEROCYTOSIS	4 patients
LEFT SIDED PORTAL HYPERTENSION	2 patients
TRAUMA	2 patients
SPLENIC TORSION	1 patient
VASCULAR MALFORMATION	1 patient
SCD	1 patient
PANCREATIC TUMOUR	1 patient
SPLENIC ABSCESS	1 patient

S. NO.	AGE	SEX	INDICATIONS	VACCINATION STATUS	AVERAGE SPLENIC WEIGHT	INTRA OPERATIVE COMPLICATIONS	POST OPERATIVE COMPLICATIONS	FOLLOW UP
1	12 YRS	F	ITP	RECEIVED	600GMS	NIL	SUPERFICIAL WOUND INFEC-TION	2 YEARS
2	11 YRS	M	THALESSEMIA	RECEIVED	410 GMS	NIL	NIL	1.5 YEARS
3	10 YRS	M	HEREDIATARY SPHEROCYTO-SIS	RECEIVED	320 GMS	NIL	NIL	2 YEARS
4	9 YRS	M	LEFT SIDED PORTAL HYPER-TENSION	RECEIVED	360 GMS	BLEEDING	NIL	2 YEARS
5	3 YRS	F	TRAUMA	RECEIVED	290 GMS	BLEEDING	NIL	1.5 YEARS
6	10 YRS	F	VASCULAR MALFORMA-TION	RECEIVED	300 GMS	BLEEDING	NIL	2 YEARS
7	12 YRS	M	SICKLE CELL DISEASE	RECEIVED	370GMS	NIL	NIL	2 YEARS
8	9 YRS	F	PANCREATIC TU- MOUR	RECEIVED	260 GMS	NIL	PNEUMOTHORAX	1 YEAR
9	5 YRS	F	SPLENIC AB-SCESS	RECEIVED	370 GMS	NIL	SUPERFICIAL WOUND INFEC-TION	2 YEARS

Discussion

The role of the spleen has long been disputed. Splenomegaly was associated with poor health and poor athletic ability in the ancient Roman, Egyptian and Babylonian eras[3].The first recorded splenectomy in Western medicine occurred in 1549, but it was not until the 19th century that elective splenectomies for splenomegaly were regularly performed. Due to advances in anatomical knowledge and surgical technique, splenectomy became increasingly feasible. Fortunately, this improvement in surgical capability was accompanied by the knowledge that splenectomy performed for the wrong indication often had disastrous consequences.

Currently, the most common indication for splenectomy in children is to treat the splenic effects of haematological conditions, e.g. hypersplenism, splenomegaly or splenic sequestration.The first splenectomy for haematological disorders in children was performed in the early 20th century, more than 400 years after the first recorded splenectomy in adults[4].

Splenectomy has been suggested as a possible therapeutic approach to manage severely affected pa-tients, based on the evidence that abnormal or damaged red blood cells passing through the spleen red pulp are removed by the splenic macrophage system. However, although splenectomy has been com-monly used in recent decades in the clinical management of patients with severe haematologic pheno-types. All these patients were managed medically first.

Children are prone to trauma due to massive splenomegaly which can be a life threatening condition.

The thalassemias are a diverse group of genetic blood diseases characterised by absent or decreased production of normal haemoglobin, resulting in a microcytic anaemia of varying degree. It is estima-ted that there are about 65,000-67,000 β-thalassemia patients in India[5]. There are two primary types of Thalassemia disease: Alpha Thalassemia disease and Beta Thalassemia disease. Proper treatment in-cludes routine blood transfusions and other therapies. Size of the spleen and frequency of transfusions are the factors on which the indication for splenectomy depend.

Immune thrombocytopenic purpura (ITP) is an acquired immune-mediated disease characterized by a decrease in platelet count due to antiplatelet autoantibody- mediated increased platelet destruction and, in some cases, associated impaired platelet production. ITP cases otherwise respond well to ster-oids. Steroid refractory cases are the indication for splenectomy[6]. Following thalassemia syndrome and sickle cell disease (SCD), hereditary spherocytosis (HS) is the most common form of congenital hemolytic anemia with an incidence of approximately 1:2000 and a dominant transmission in about 70–80% of cases. HS is caused by mutations in genes encoding α-and β-spectrin and other proteins involved in the attachment of the cytoskeleton to the overlying lipid bilayer (ankyrin, band 3 and protein 4.2). Defects in these structural proteins render the red blood cells spherical, rigid and susceptible to premature destruction in the spleen. Congenital dyserythro-poietic anemia (CDA) is a group of rare red blood cell disorders characterized by ineffective erythro-poiesis, pathognomonic cytopathology of nucleated red blood cells in bone marrow and increased iron absorption with secondary hemochromatosis . SCD is caused by a point mutation in the β-globin gene resulting in the synthesis of a pathological hemoglobin, HbS.90 Cyclic polymerization/depoly-merization of deoxy-HbS generates dense, dehydrated red cells that play a central role in the acute and chronic clinical manifestations of SCD, in which intravascular sickling leads to vaso-occlusion and impaired blood flow with ischemic/reperfusion injury[7].

The main cause of Left Sided Portal Hypertension (LSPH) is splenic vein thrombosis (SVT). Acute and chronic pancreatitis and pancreas neoplasms are the most common causes of SVT. Splenomegaly is a hallmark of long-standing portal hypertension and is frequently seen in patients with LSPH. A pa-tient with active bleeding unresponsive to conservative management should be operated on quickly. Splenectomy is the treatment of choice.

Wandering spleen (WS) is a clinical entity which rarely affects children and adolescents. It is caused by laxity or absence of the sup-porting splenic ligaments, allowing the spleen to be mobile within the abdomen and predisposing to the torsion along the vascular pedicle. It appears as a surgical emergency when an acute twisting occurs. In case of acute torsion, most authors prefer to preserve the spleen and perform a splenopexy.

The spleen is the most commonly injured intra-abdominal organ in children [8]. Compared to adults, the abdominal organs in children are at a higher risk of organ lesions due to a higher transmission rate of forces through the thinner abdominal wall, larger relative surfaces of the spleen and liver, more flexible ribs and the more horizontal positioning of the diaphragm in children compared to adults.

Splenic haemangiomas, also known as splenic venous malformations, splenic cavernous malforma-tions, or splenic slow flow venous malformations, while being rare lesions, are considered the second commonest focal lesion involving the spleen after simple splenic cysts and the most common primary benign neoplasm of the spleen.

Spleniculi or accessory spleen, is a congenital anomaly occurring in 10% of individuals.

Spleniculi may be solitary or multiple, seldom >6 and are more commonly involved in hematological disorders of the spleen. Common sites of occurrence include the splenic hilum (75%) and the 2nd most common being near pancreatic tail (20%). Accessary spleen can enlarge following splenect-omy and may be source of recurrent symptoms.

Autologous blood transfusion is the collection of blood from a single patient and retransfusion back to the same patient when required. The primary driving forces for the use of autologous blood transfu-sion are to reduce the risk of transmission of infection and to protect an increasingly scarce resource. There were 2 patients in our setup who underwent autologous blood transfusion.

Notwithstanding significant improvements in the post-operative haematological profiles of these pa-tients, splenectomised patients were also found to be especially prone to immediate and late postoperative sepsis. By the 1970s, this phenomenon and the associated role of encapsulated bacteria, such as S. pneumoniae, H. influenzae type B and Neisseria meningitides types A and C, had been labelled as overwhelming causes of post splenectomy infection (OPSI). OPSI is a devastating consequence of splenectomy in children, with mortality rates of up to 50% in high-risk groups, i.e. young children, those with haematological disease[9]. The risk of OPSI is lifelong. Appropriate pre- and postoperative management with vaccina-tion, antibiotic prophylaxis and immediate management of suspected sepsis in splenectomised patients have enabled modern surgeons and physicians to safely perform splenectomies in children. In our setup, oral PENTID is given as antibio-tic prophylaxis. The indications for splenectomy in our series were similar to those found in the international litera-ture, with the majority of cases performed for haematological disorders (namely Thalassemia, ITP and HS).

The complication rate for open splenectomy was 9%, with major complications accounting for 4.5% and minor complications 4.5% of the overall rate. This finding may be due to numerous factors including the severity of the underlying disease process at the time of surgery and may suggest that earlier splenectomy may decrease the complication rate in our setting. Within this short span of period, no mortality was recorded. No case of OPSI was recorded.

Table 2: Complications

1.	MINOR COMPLICATIONS	
	- Superficial wound sepsis	1
2.	MAJOR COMPLICATIONS	
	-Right Pneumothorax	1
3.	PVT	NIL
4.	OPSI	NIL

Conclusion

Indications for splenectomy were similar locally to those noted internationally, with haematological disorders accounting for the majority of cases. Although no patients experienced OPSI in our series.

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