

Pediatric surgery experiences of a tertiary referral hospital: International Classification of Diseases spectrum for teaching, planning, and scaling up services

Col Sunil Jain¹, Lt Col Naveen Chandra², Col Rajeev Kumar Thapar³

From, Classified Specialist, Departments of ¹Paediatrics, ²Paediatric Surgery, ³Senior Advisor, Department of Paediatrics, Command Hospital, Lucknow, Uttar Pradesh

Correspondence to: Col Sunil Jain, Department of Paediatrics, Command Hospital (NC), Pin 901131, c/o 56 APO, India. E-mail: sunil_jain700@rediff.com

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ABSTRACT

Introduction: Pediatric surgery provides the opportunity to intervene positively in a wide array of diseases with potential lifelong impact. **Objective:** The objective of this study was to evaluate the significance of pediatric surgery under vast circumstances. **Materials and Methods:** All children in the age group of 0–10 years operated in a tertiary care hospital were analyzed. Grouping of cases was done using the International Statistical Classification of Diseases and Related Health Problems 10th Revision (International Classification of Diseases [ICD]-10)-WHO Version (2016) and incorporating terminology from the ICD-11 Beta Draft. **Results:** “Developmental anomalies” accounted for 79.1% of cases, while 20.9% were “acquired conditions requiring surgical intervention.” The common congenital malformations were those of the genital organs (17%), followed by those of the digestive (13%) and nervous system (13%), urinary system (12%), circulatory system (8%), and cleft lip and palate (7%). The essential surgery package for congenital anomalies should include expertise in repair of cleft lip and palate, anomalies of genital, digestive, nervous, urinary, and musculoskeletal system. Referral to superspecialty center is required only for “congenital malformations of the circulatory system.” **Conclusion:** Majority of the workload (79.1%) due to “developmental anomalies” points toward need for skills in correcting these, restoring anatomy for the proper growth, and physiological functioning. Inclusion of indirect inguinal hernia in the Chapter 20 ‘Developmental anomalies’ (ICD-11) will contribute to correct international comparisons and guide planning.

Key words: *Congenital anomalies, Indirect hernia, Pediatric Surgery*

Children are not small adults and pediatric surgery requires the greatest sophistication and scientific skills. Pediatric surgery provides the opportunity to intervene positively in a wide array of diseases [1], with potential lifelong impact, hence, the need for tactful evaluation for thoughtful evidence of pediatric surgery. The essential surgery package for congenital anomalies at the second- and third-level hospitals includes (i) repair of cleft lip and palate, (ii) repair of club foot, (iii) shunt for hydrocephalus, and (iv) repair of anorectal malformations and Hirschsprung’s disease [2]. Importantly, it has been suggested that scaling up selected subspecialty surgical care can contribute significantly in preventing mortality. The largest chunks of these preventable deaths are due to congenital heart anomalies (66%) and neural tube defects (17%) [3]. Amidst all this thinking, it becomes important to systematically analyze the features of & facilities for Paediatric surgery at a referral hospital.

The International Statistical Classification of Diseases and Related Health Problems (International Classification of Diseases [ICD]) is the foundation for the identification of health trends and statistics globally, and the international standard for reporting diseases and health conditions. It is the

diagnostic classification standard for all clinical and research purposes. Pediatric surgical conditions classification as per the ICD can definitely add to teaching systematically and planning purposes.

MATERIALS AND METHODS

All children admitted for surgery in the age group of 0–10 years over a period of 1 year from January to December 2017 were studied. The setting was a tertiary care referral hospital of the Indian Armed Forces located at Uttar Pradesh, India. Total numbers of the cases operated were 225 during the study period and these were included in the study. Cases not operated were excluded from the study. Data of diagnosis, surgery performed, and complications/outcome, collected from the hospital registry, were analyzed.

Grouping of cases was done using the ICD-10-WHO Version (2016) and incorporating terminology from the ICD-11 Beta Draft. For spectrum of conditions, the percentage of various ICD blocks was calculated. This being a descriptive study, analyzing the number of cases operated as ICD spectrum, only simple statistics used as percentage of cases. No ethical issues involved in

the study, as only retrospective data analysis of diagnosis, surgery done, and complications/outcome carried out from the hospital records. No revelations of any identifiable human material and data being done.

RESULTS

A total of 225 cases were operated during the study period and were included in the study. The different pediatric surgical conditions dealt, surgical intervention done, complications, if any and outcome are given in Tables 1-10 for Chapter 20 “developmental anomalies” (ICD-11 Beta Draft)/XVII “congenital malformations, deformations, and chromosomal abnormalities” (ICD-10) and Table 11 for “acquired conditions requiring surgical intervention.” The total number of cases dealt with is given in each table. Comprehensively, Fig. 1 gives the graphical depiction of various groups.

DISCUSSION

Pediatric surgeons are challenged with the assessment and organization of a wide spectrum of surgical pathologic processes ranging from head and neck lesions to thoracic and gastrointestinal tract anomalies, oncologic disorders, and trauma [4]. Systematic analysis for sophisticated advancements is regularly required. William Farr had stated in 1856, “classification is a method of generalization. Several classifications may, therefore, be used... to facilitate inquiries, and to yield general results” [5]. Our study used the ICD for uniformity and international comparisons. The ICD Chapter 20 “developmental anomalies” (ICD-11 Beta Draft)/XVII “congenital malformations, deformations, and chromosomal abnormalities” (ICD-10) accounted for 79.1% of cases, while “acquired conditions requiring surgical intervention” were 20.9%. This points toward the need for skills for congenital

Table 1: Congenital malformations of the nervous system (ICD Code Q00-Q07), (n=30)

ICD classification	n	Surgical intervention	Complications/outcome
Q01 – Encephalocele			
Q01 Meningocele, cerebral	01	Excision of herniated tissue and closure of defect	Nil/progressing well
Q01.2 Occipital encephalocele	01	Excision of herniated tissue and closure of defect	Nil/on follow-up for vision problems
Q03 – Congenital hydrocephalus			
Q03.1 Congenital hydrocephalus - Atresia of foramina of Magendie and Luschka (Dandy-Walker syndrome)	02	VP shunt insertion	Shunt revision 01/On follow-up for cerebellar ataxia, and delayed motor and cognitive milestones
Q03.9 Congenital hydrocephalus, unspecified	07	VP shunt insertion	Shunt revision 04/on follow-up for delayed developmental
Q05 – Spina bifida			
Q05.2 Lumbar spina bifida with hydrocephalus	04	Surgical closure with VP shunt insertion	Shunt revision 02
Q05.5 Cervical spina bifida without hydrocephalus	01	Surgical closure with VP shunt insertion	Nil/progressing well
Q05.7 Lumbar spina bifida without hydrocephalus	08	Surgical closure	Nil/on follow-up
Q05.8 Sacral spina bifida without hydrocephalus	04	Surgical closure	Nil/on follow-up
Q07 – Other congenital malformations of nervous system			
Q07.0 Arnold-Chiari syndrome	02	Surgical decompression 01/conservative management 1	Nil/on follow-up

ICD: International Classification of Diseases, VP: Ventriculoperitoneal

Table 2: Congenital malformations of eye, ear, face, and neck (ICD Code Q10-Q18), (n=10)

ICD classification	n	Surgical intervention	Complications/outcome
Q10 – Congenital malformations of eyelid, lacrimal apparatus, and orbit			
Q10.0 Congenital ptosis	02	Levator resection 01, frontalis suspension procedure 01.	Nil/on follow-up
Q10.2 Congenital entropion	01	Surgical correction	Nil/favorable
Q12 – Congenital lens malformation			
Q12.0 Congenital cataract	03	Surgical removal of lens material	Nil/correction of resultant aphakic refractive error
Q16 – Congenital malformations of ear causing impairment of hearing			
Q16.1 Congenital absence, atresia, and stricture of auditory canal (external)	02	Reconstructive surgery	Nil/favorable
Q17 – Other congenital malformations of ear			
Q17.2 Microtia	02	Reconstructive surgery	Nil/favorable

ICD: International Classification of Diseases

Table 3: Congenital malformations of the circulatory system (Q20-Q28) (n=17)

ICD classification	n	Surgical intervention	Complication/outcome
Q20 Congenital malformations of cardiac chambers and connections			
Q20.0 Common arterial trunk – Persistent truncus arteriosus	01	Referred to CTS center, corrective surgery done	Nil/on follow-up
Q20.3 Discordant ventriculoarterial connection – Transposition of great vessels (complete)	01	Referred to CTS center, arterial switch (Jatene) procedure done	Nil/on follow-up
Q21 Congenital malformations of cardiac septa			
Q21.3 Tetralogy of Fallot	03	Referred to CTS center, corrective surgery done	Nil/on follow-up
Q22 Congenital malformations of pulmonary and tricuspid valves			
Q22.1 Congenital pulmonary valve stenosis	01	Balloon valvuloplasty done	Nil/on follow-up
Q22.5 Ebstein anomaly	01	Referred to CTS center, Starnes operation done	Nil/on follow-up
Q25 Congenital malformations of great arteries			
Q25.0 Patent ductus arteriosus	06	04 managed medically/02 referred to CTS center	Nil/on follow-up
Q25.1 Coarctation of aorta	03	Referred to CTS center, surgical repair done	Nil/on follow-up
Q25 Congenital malformations of great veins			
Q26.3 Partial anomalous pulmonary venous connection	01	Referred to CTS center, surgical correction done	Nil/on follow-up

CTS: Cardiothoracic surgery, ICD: International Classification of Diseases

Table 4: Congenital malformations of the respiratory system (Q30-Q34), (n=01)

ICD classification	n	Surgical intervention	Outcome
Q34 – Other congenital malformations of respiratory system			
Q34.1 Congenital cyst of mediastinum	01	Excision	On follow-up

ICD: International Classification of Diseases

Table 5: Cleft lip and cleft palate (Q35-Q37), (n=15)

ICD classification	n	Surgical intervention	Complications/outcome
Q35 cleft palate			
Q35.1 Cleft hard palate	01	Surgical repair	Nil/on follow-up
Q35.3 Cleft soft palate	02		
Q35.5 Cleft hard palate with cleft soft palate	03		
Q36 Cleft lip			
Q36.0 Cleft lip, bilateral	02	Surgical repair	Nil/on follow-up
Q36.1 Cleft lip, median	01		
Q36.9 Cleft lip, unilateral	02		
Q37 Cleft palate with cleft lip			
Q37.0 Cleft hard palate with bilateral cleft lip	02	Surgical repair	Nil/on follow-up
Q37.1 Cleft hard palate with unilateral cleft lip	02		

ICD: International Classification of Diseases

conditions, in correcting these, restoring anatomy for proper growth and physiological functioning.

The ICD Block “congenital malformations of genital organs” accounted for the maximum number of cases (16.4%). Hypospadias and undescended testicle accounted the maximum number of cases. The incidence of hypospadias appears to be increasing [6], hence, the need for requisite surgical skills. Timely surgical intervention for undescended testicle surgery has the advantages that early scrotal placement reduces the risk of malignancy, infertility, and torsion. It improves the endocrine function of the testis and creates a normal-appearing scrotum. The complications after surgery are low, although atrophy and retraction of testis and post-operative hernia are possibilities.

“Congenital malformations of the nervous system” were the second with 13.3% of total cases operated. Spina bifida and

congenital hydrocephalus were the most common in this ICD block. Myelomeningocele is the most common type of spina bifida aperta (also lately and commonly referred as Manifesta). Open myelomeningocele presents a high infection risk and requires early surgical repair. All cases in our setup were treated with ventriculoperitoneal shunt insertion. Shunt revision rates were high (55.5%). Endoscopic third ventriculostomy has emerged as an advantageous and appealing alternative for an obstruction at the aqueduct or the outlet of the fourth ventricle and thus the need for expediting the expertise and equipment for this.

The third common ICD block in our study was “other congenital malformations of the digestive system” accounting for 13% of the cases. Conditions vary from those from the oral cavity to anorectal malformations (ARM). ARMs are among the more frequent congenital anomalies encountered in pediatric

Table 6: Other congenital malformations of the digestive system (Q38-Q45), (n=29)

ICD classification	n	Surgical intervention	Complications/outcome
Q38 – Other congenital malformations of tongue, mouth, and pharynx			
Q38.1 Ankyloglossia, tongue tie	14	Surgical excision	Nil/favorable
Q40 – Other congenital malformations of upper alimentary tract			
Q40.0 Congenital hypertrophic pyloric stenosis	03	Pyloromyotomy	Nil/favorable
Q41 – Congenital absence, atresia, and stenosis of small intestine			
Q41.0 Congenital absence, atresia, and stenosis of duodenum	02	Duodenoduodenostomy	Nil/anastomotic leak (01)
Q42 – Congenital absence, atresia, and stenosis of large intestine			
Q42.0 Congenital absence, atresia, and stenosis of rectum with fistula	02	Posterior sagittal anorectoplasty	Nil/fecal incontinence (1)
Q42.2 Congenital absence, atresia, and stenosis of anus with fistula	02	Cutback anoplasty 01 Transposition anoplasty 01	Nil/on follow-up
Q42.3 Congenital absence, atresia, and stenosis of anus without fistula imperforate anus	01	Serial dilation	Nil/on follow-up
Q43 – Other congenital malformations of intestine			
Q43.0 Meckel diverticulum	01	Wedge resection of diverticulum with transverse closure of ileum	Nil/favorable
Q43.1 Hirschsprung disease	02	Endorectal pull-through	Nil/on follow-up
Q43.5 Ectopic anus	02	Cutback anoplasty 01 Transposition anoplasty 01	Nil/on follow-up

ICD: International Classification of Diseases

Table 7: Congenital malformations of genital organs (Q50-Q56), (n=37)

ICD classification	n	Surgical intervention	Complications/outcome
Q52 – Other congenital malformations of female genitalia			
Q52.3 Imperforate hymen	02	Hymenotomy	Nil/favorable
Q53 – Undescended testicle			
Q53.0 Ectopic testis	03	Laparoscopy	Nil/on follow-up
Q53.1 Undescended testicle, unilateral	06	Standard Dartos pouch orchidopexy – 03, Open inguinal orchidopexy – 02, Laparoscopic orchidopexy – 01	Nil/on follow-up
Q53.2 Undescended testicle, bilateral	03	Laparoscopy two-stage Fowler-Stephens – 01	Nil/on follow-up
Q54 – Hypospadias			
Q54.0 Hypospadias, balanic glandular	02	Meatal advancement and glansplasty	Nil/on follow-up
Q54.1 Hypospadias, penile	13	Tubularized incised plate urethroplasty	Meatal stenosis 01
Q54.2 Hypospadias, penoscrotal	08	Two-stage repair	Fistula 1 Meatal stenosis 1

ICD: International Classification of Diseases

Table 8: Congenital malformations of the urinary system (Q60-Q64), (n=28)

ICD classification	n	Surgical intervention	Complications/outcome
Q62 – Congenital obstructive defects of renal pelvis and congenital malformations of ureter			
Q62.0 Congenital hydronephrosis	07	Dismembered pyeloplasty	Nil/on follow-up
Q62.3 Other obstructive defects of renal pelvis and ureter – Congenital ureterocele	02	Laparoscopic excision of the obstructed upper pole and most of the associated ureter – 01 Transurethral incision with cautery, f/b subsequent excision of ureterocele and ureteral reimplantation – 01	Nil/on follow-up
Q62.7 Congenital vesicoureterorenal reflux	11	Ureteral reimplantation	Nil/on follow-up
Q64 – Other congenital malformations of urinary system			
Q64.1 Exstrophy of urinary bladder – Ectopia vesicae	01	Staged repair	Nil/on follow-up
Q64.2 Congenital posterior urethral valves	07	Transurethral ablation	Nil

ICD: International Classification of Diseases

surgery [7]. This complicated and challenging group represents a wide spectrum of defects. We managed three cases of anorectal malformation, two with low lesions and one with high lesion. As

per the latest Krickenbeck classification of ARM, all the cases were of major clinical groups. The first two were males with perineal (cutaneous) fistula. Both were operated with cutback

Table 9: Congenital malformations and deformations of the musculoskeletal system (Q65-Q79), (n=07)

ICD classification	n	Surgical intervention	Complications/outcome
Q68 – Other congenital musculoskeletal deformities			
Q68.0 Congenital deformity of sternocleidomastoid – Contracture of sternocleidomastoid (muscle)	02	Transection	Nil
Q79 Congenital malformations of the musculoskeletal system, not elsewhere classified			
Q79.0 Congenital diaphragmatic hernia	03	Open surgical repair	Survived 01
Q79.2 Exomphalos, Omphalocele	01	Staged closure	Nil
Q79.3 Gastroschisis	01	Staged repair	Nil

ICD: International Classification of Diseases

Table 10: Other congenital malformations (Q80-Q89), (n=04)

ICD classification	n	Surgical intervention	Complications/outcome
Q85 – Phakomatoses, not elsewhere classified			
Q85.0 Neurofibromatosis (non-malignant) Von Recklinghausen disease, Plexiform	02	Resection	Repeat resection
Q89 Other Congenital malformations, not elsewhere classified			
Q89.2 Congenital malformations of other endocrine glands – Persistent thyroglossal duct	02	Resection	Nil

ICD: International Classification of Diseases

Table 11: Acquired conditions requiring surgical intervention, (n=47)

ICD classification	n	Surgical intervention	Complications
Chapter VI diseases of the nervous system (G00-G99)			
G91.0 Communicating hydrocephalus	02	VP shunt insertion	Nil
Chapter VIII diseases of the ear and mastoid process (H60-H95)			
H66.2 Chronic atticofuruncular otitis media	03	Tympanomastoidectomy	Nil
Chapter XI diseases of the digestive system (K00-K93)			
K35.3 acute appendicitis with localized peritonitis	07	Appendicectomy	Nil
K80.2 Calculus of gallbladder without cholecystitis	06	Laparoscopic cholecystectomy	Nil
K40.2 Bilateral inguinal hernia, without obstruction or gangrene	04	Open repair 02 Laparoscopic repair 02	Nil
K40.9 Unilateral or unspecified inguinal hernia, without obstruction or gangrene	18	Open repair 11 Laparoscopic repair 07	Nil
K41.9 Unilateral or unspecified femoral hernia, without obstruction or gangrene	01	Surgical repair	Nil
Chapter XIV Diseases of genitourinary system (N00-N99)			
N20.0 Calculus of kidney	04	Extracorporeal shock wave lithotripsy 03 Percutaneous nephrostolithotomy 01	Nil
N20.1 Calculus of ureter	02	Extracorporeal shock wave lithotripsy 02	Nil

ICD: International Classification Of Diseases

anoplasty. The third was female, a Down's syndrome case, and high ARM with no fistula. Three-stage repair was done for this case, initial diverting colostomy followed by limited posterior sagittal anorectoplasty (PSARP), and finally, colostomy closure was performed.

At present, two main approaches are used for reconstruction of ARM. The first is the PSARP and the second is the laparoscopy-assisted anorectoplasty. We used the PSARP as laparoscopic approach has the potential to leave behind a small diverticulum at the site of the fistulous connection [8].

The next and important ICD block was "congenital malformations of the urinary system" (12.4%). Many children with congenital hydronephrosis due to an apparent pelvi-ureteral obstruction are not highly obstructed and will improve with time. However, patients with infections or impaired renal function require repair to improve drainage. Open dismembered

pyeloplasty is considered the gold standard approach, especially in infants. In older children, laparoscopic or robotic approaches for pyeloplasty can expedite convalescence and diminish post-operative pain. An endoscopic approach, endopyelotomy, is also an option in older children. Posterior urethral valves are the most common cause of severe obstructive uropathy in children. Approximately 30% of patients experience end-stage renal disease or chronic renal insufficiency [9]; hence, there is a need of the prompt attention and surgical intervention.

In our study, we had 17 cases of "congenital malformations of the circulatory system." All, except one, required referral to a cardiothoracic surgery (CTS) center. We had no mortality with early diagnosis and proper transport to CTS center. Congenital heart diseases which are operated in neonatal period are TGA, TOF, CoA, PDA, TAPVC, and Truncus arteriosus, while palliative surgery is carried out for hypoplastic left heart syndrome and

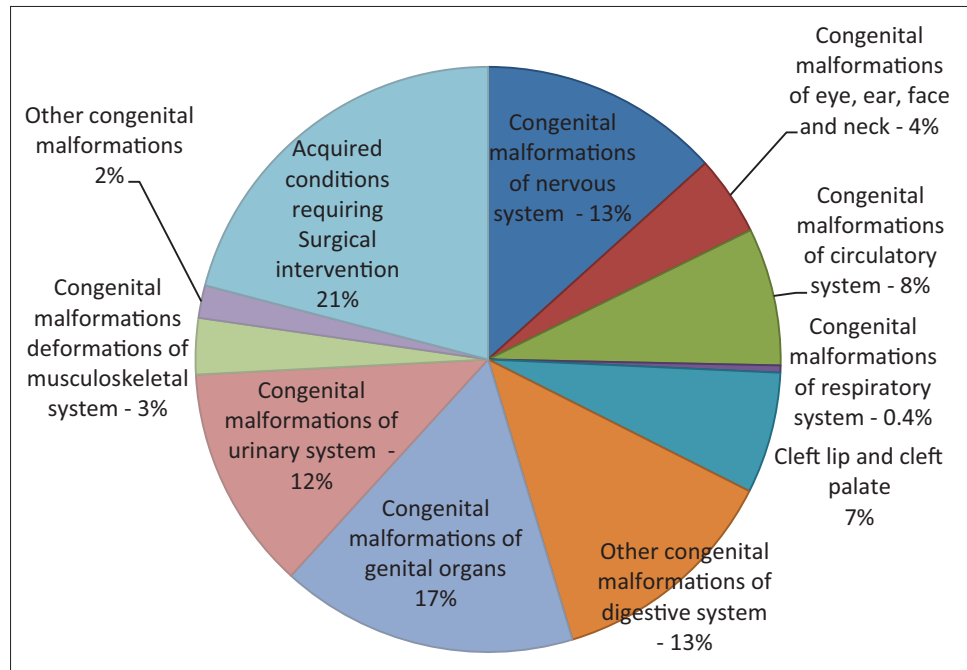


Figure 1: Pediatric surgical conditions

complex lesions [10]. The largest chunk of preventable deaths for want of subspecialty surgical care is due to congenital heart anomalies (66%) [3]. Incidence as well as expertise requirements justify the need for and/or facilities for transport to pediatric cardiac surgery center.

“Cleft lip and cleft palate” cases were only 6.70% in our study. It has been suggested that the essential surgery package for congenital anomalies at the second- and third-level hospitals should include repair of cleft lip and palate [2]. Incidence varies between races from 1:300 to 1:2000 live births [11]; hence, resources should be allocated as per local incidence. All children born with a cleft lip and palate need a thorough pediatric assessment to exclude other congenital abnormalities [11]. Hence, we suggest strategy of prompt assessment by pediatricians and referral as required.

The incidence of the ICD block “congenital malformations of eye, ear, face, and neck” is low (4.4%), but diversity justifies multifaceted/subspecialty expertise. The next ICD block is congenital malformations and deformations of the musculoskeletal system with 3.1% of cases. We had three cases of congenital diaphragmatic hernia. One case of CDH survived with surgery. The other two could not be salvaged due to pulmonary hypoplasia in one and associated large VSD in the other. In cases of CDH, diligent initial stabilization is important. This requires aggressive respiratory support. Relative predictors of a poor prognosis, as in our two cases which could not be salvaged, are associated major anomaly and severe pulmonary hypoplasia [12]. In the ICD block, “Other congenital malformations” cases, only 1.80%, were managed with resection.

Acquired conditions requiring surgical intervention were 20.9% and diverse. Abdominal symptoms are possibly the most frequent of all symptoms encountered in surgical practice [13]. Similar is the incidence in pediatric surgical practice with 36 cases

of total 47 in the acquired conditions from the ICD Chapter “diseases of the digestive system” in our study.

The ICD classification needs modification with regard to inguinal hernias, for a better reasoning based representation as follows: (i) The overwhelming majority of inguinal hernias in infants and children are congenital indirect hernias (99%) as a consequence of a patent processus vaginalis, a developmental structure important in testicular descent and (ii) approximately 50% of inguinal hernias manifest clinically in the 1st year of life, most in the first 6 months [14]. The incidence, the age of presentation, and the congenital origin as a result of anomalous patency of developmental structure justify inclusion of indirect inguinal hernia in Chapter 20 “developmental anomalies” (ICD-11 Beta Draft)/Chapter XVII “congenital malformations, deformations, and chromosomal abnormalities” (ICD-10).

Pediatric surgery is the last bastion of a true general surgical specialty; delivering comprehensive surgical care covering a broad scope of conditions in infants, children, and young adults [4]. Our study reiterates that “*Pediatric surgery is a true general specialty that is distinctive, delivering comprehensively with diligence, covering a wide spectrum of conditions with sophistication for success.*”

CONCLUSION

In our study, 79.1% of the cases had congenital anomalies and the rest had acquired conditions. Congenital malformations of the genital organs were the most common disease followed by those of digestive, nervous, urinary system, cleft lip and palate, and those of musculoskeletal system. The essential surgery package for congenital anomalies at the second- and third-level hospitals should include expertise for anomalies of all systems. Referral to superspecialty center was required for only “congenital

malformations of the circulatory system.” Incidence as well as expertise requirements justify the need for and/or facilities for transport to pediatric cardiac surgery center and can prevent mortality.

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