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Prevalence of some rare pediatric diseases, their complications and treatment analysis

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Abstract

The study was aimed to evaluate the prevalence of some rare pediatric diseases; mainly hypospadias, hernia and cystic fibrosis along with their complications and treatment analysis. The major objective of the study includes demographic analysis, evaluation of diseases and analysis of treatment. The study was carried out on both in and out patients of different hospitals/clinics or nursing homes for collection of data. A total number of 175 pediatric patients were the part of the study and out of 175 patients, male children's in the study were 74.86%, while female children's were 25.14% of the study population. Study showed that patients of hernia were more as compared to cystic fibrosis and hypospadias which reflected that hernias are more common in patients as compared to other two diseases. The study also showed that majority of patients in all age group was males as compared to females. It was also analyzed from the study that hypospadias, a disease that occurs mostly in males, is a problem of concern in the society because of its complicated pathology as well as tedious approach of treatment. It was suggested that being critical problems of concern, such diseases and their diagnosis, care, prevention and management need to be look after seriously in the field of medicine and such studies could play a beneficial role for healthcare practitioners as well as for the society.

Keywords: Pediatric, hypospadias, hernia, cystic fibrosis, prevalence

Introduction

Pediatrics is the branch of medicine that mainly deals with the diagnosis, prevention, treatment or cure of diseases in infants, children's and adolescents. Diseases are always devastating for anyone, but it seems unfair when they attack infants or children. Children are more prone to diseases for a number of different reasons. The major reason for their increased susceptibility to diseases is that they haven't yet built immunologic defenses required to fend off certain diseases^[1,2].

Hypospadias^[2,3]

Hypospadias is a common birth defect of the penis in which the urethra is not at the tip of the penis resulting in incomplete development of the anterior urethra. It is characterized by the abnormal urethral opening on the ventral shaft of the penis. It can be located on the undersurface of the penis and structures affected in various degrees of deficiency of the urethra, corpora cavernosa and corpus spongiosum. The skin on the ventral surface may be thin, and the prepuce is deficient ventrally and forms a dorsal hood over the glans. A condition called chordee is often seen with hypospadias. It is an abnormal downward curve of the penis that can occur with or without a hypospadias. Hypospadias is defined as the combination of three anatomical abnormalities of the penis:

1. Proximal urethra, with or without ectopic stenotic meatus.
2. A ventral penile shaft deviation
3. A typical dorsal winged prepuce.

Hernia^[4-6]

A hernia is the prominence of an organ through the wall of the cavity that normally contains it. There are many different kinds of hernias, requiring specific management or treatment with respect to specific case. The most common hernia develop in the abdomen, when a weakness in the abdomen wall develops into a localized hole or defect through which abdominal organs or adipose tissues covered with peritoneum, may protrude. Another common type of hernia involves the spinal discs and causes sciatica. A hiatus hernia is the type of hernia that occurs when the stomach protrudes through the esophageal opening in the diaphragm.

Causes of hiatus hernia depend on each individual. Among the multiple causes, mechanical causes include: improper heavy weight lifting, hard coughing bouts, sharp blows to the abdomen, and incorrect posture. Further, conditions that increase the pressure of the abdominal cavity may cause hernias or may also worsen the existing condition. Inguinal hernias are the most common type of hernia in both men and women. In some selected cases, they may require surgery. Femoral hernias occur just below the inguinal ligament, when abdominal contents pass into the weak area at the posterior wall of the femoral canal. In hiatus hernia, the normal passageway through which the esophagus meets the stomach serves as a functional defect, allowing part of the stomach to herniate into the chest. A congenital hernia, occurring in up to 1 in 2000 births, requires pediatric surgery. Intestinal organs may herniate through several parts of the diaphragm. Hernias can be classified according to their anatomical location that includes: abdominal hernia, diaphragmatic hernias, hiatus hernias, anal hernias etc.

Cystic Fibrosis [7]

Cystic fibrosis is a genetic disorder that affects mostly lungs and occurs due to mutation of the gene for cystic fibrosis transmembrane conductance regulator (CFTR). CFTR is involved in production of sweat, digestive fluid and mucus. Cystic fibrosis is most common among northern European and least common in Africans and Asian. Cystic fibrosis, also known as mucoviscidosis, is a genetic disorder that affects mostly the lungs but also the pancreas, liver, kidneys and intestine. Long term issues include difficulty breathing and coughing up sputum as a result of frequent lung infections. Other symptoms include sinus infections, poor growth, fatty stool, clubbing of the finger and toes, and infertility in males among others. Different people may have different degrees of symptoms. There is no cure for cystic fibrosis. Lung infections are treated with antibiotics which may be given intravenously, inhaled, or by mouth. Sometimes the antibiotic azithromycin is used long term. Inhaled hypertonic saline and salbutamol may also be useful. Lung transplantation may be an option if lung function continues to worsen.

The aim of the study was to evaluate the prevalence of these rare pediatric diseases, complications and treatment analysis. The main objective of the study includes data collection of major pediatric diseases, overall demographic analysis, evaluation of diseases, analysis of treatment and any adverse drug reactions or drug interactions reported during therapy. These pediatric diseases are not common and one of the major work was to aware the general public or parents about these diseases or disorders, their treatment and prevention. Such kind of study could play a beneficial role for healthcare practitioners as well as for the society.

Methodology

Study Design

Open and prospective study: In this study, the data were collected at multiple stages or intervals as the patient's treatment undergo for long time. Here, we collected the data on the present medical status and then followed it into the future in order to record the development of any outcome. The follow up with the patient's guardian was done by mail questionnaires, by telephonic interview, via an internet, verbal communication.

Prescription and current medical records: Patient's related information, drug therapy and other relevant information were needed for the study. Such information's were taken from the patient's prescription and current medical records. Prescription helps to detect what medicines were prescribed by the doctor or specialist. Current medical records help to collect the various information's about the patient that includes patient laboratory data, diagnostic report and current prescription.

Data were collected from prescription based format and other patient related records: From the prescription based format, various information about the patient like age, gender, address and contact number were collected. The medication history of patient was collected by asking their relatives to show past medical report or past medication history.

Study Sites

The study was carried out in different hospitals/clinics or nursing homes for collection of data with appropriate prior permission from the authorized person. The selected places for study were:

- a. Suri Nursing Home;
- b. Sharda Nursing Home;
- c. Doon Hospital and
- d. Vaish Nursing Home

Study Setting

The study was proposed to be carried out on both in and out patients.

Data Sources

All the necessary and relevant information was collected through:

- In and outpatient prescription records
- Patient medication history
- Patient pathological and laboratory data reports
- Patient diagnostic reports
- Verbal communication with the patient's guardian

Results and Discussion

Data were collected from prescription based format and other patient related records obtained after confirmation of the diagnosis of the diseases by the physician. A total number of 175 pediatric patients were the part of the study. It was analyzed that, out of 175 patients, the total number of the male children's in the study were 131 accounting for 74.86% of the study population, while female children's were 44 accounting for 25.14% of the study population (Figure-1). This observation showed that gender-wise distribution of patients was more in males as compared to females. By viewing such distribution, it was observed that these diseases were more likely to occur in males children's as compared to females. One of the main reasons behind more number of male children's was the cases of hypospadias disorder that come under the category of males only. The other diseases like hernia and cystic fibrosis, their occurrence in males and females is independent of gender.

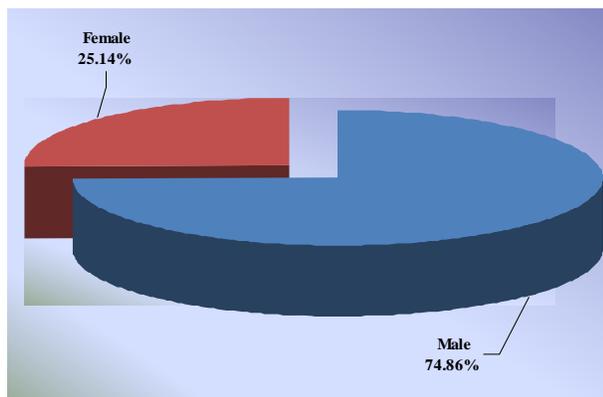


Fig 1: Gender-wise distribution of pediatric patients

Out of 175 patients, 61 patients were from age group of 2-5 years, 39 were from age group of infants, 35 were from age group of 6-10 years, 25 were from age group of 11-15 years, 15 were from age group >15 years. The study showed that majority of the patients were between 2-5 years of age and age-wise distribution was found higher in male patients than female patients (Table-1 and Figure-2). The study also showed that majority of patients in all age group was males as compared to females. One of the main reasons of high percentage of male patients was that, in hypospadias disease all patients were under the category of male and it is the disorder which occurs only in male patients.

Table 1: Distribution of patients according to age group

S. No.	Age Group	Number of Patients (%)		Total No. of Patients (%) (n=175)
		Male	Female	
1	Infant	27(15.40)	12(6.85)	39 (22.28)
2	2-5	47(26.85)	14(8.00)	61 (34.85)
3	6-10	26(14.85)	9(5.14)	35 (20.00)
4	11-15	19(10.85)	6(3.42)	25 (14.28)
5	>15	12(6.85)	3(1.71)	15 (8.75)

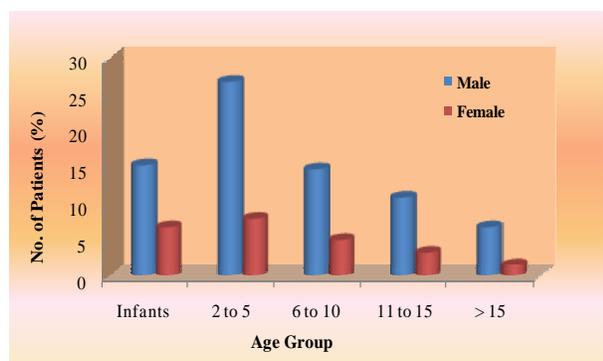


Fig 2: Distribution of patients according to age group

In this study, parents having monthly income less than 8000 rupees were considered under lower socio-economic class. For middle class, monthly income ranged from 8000 to 16000

rupees while upper class was considered where monthly income was found more than 16000 rupees. Based on the above socioeconomic distribution, the study showed that 31.42% male patients and 7.42% female patients belongs to lower class status, 24.57% male patients and 8.57% female patients belongs to middle class status, 18.85% male patients and 9.14% female patients belongs to upper class status. The greater number of patients belonged to lower class as compared to middle and upper class (Table-2 and Figure-3). From these results, it was analyzed that lower and middle class of people are more prone to these diseases as compared to upper class.

Table 2: Socio-economic distribution of patients

S. No.	Social Status	Monthly Income (Rs.)	No. of Patients (%)		Total No. of Patients (%) (n=175)
			Male	Female	
1	Lower class	<8000	55 (31.42)	13 (7.42)	68 (38.85)
2	Middle class	8000-16000	43 (24.57)	15 (8.57)	58 (33.14)
3	Upper class	>16000	33 (18.85)	16 (9.14)	49 (28.00)

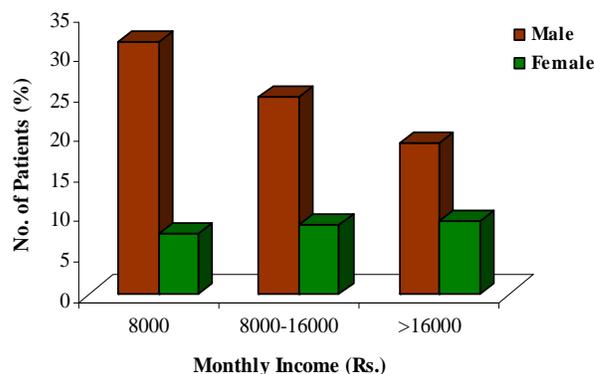


Fig 3: Socio-economic distributions of patients

It was observed that majority of infants and children's were more susceptible to diseases because of poorly developed immunologic system. In this study, diseases under study were hypospadias, hernia and cystic fibrosis. The distribution of total number of pediatric patients according to selected disease conditions was tabulated in (Table-3). The pediatric diseases under the study were evaluated based on the demographics involvement along with the cause and nature of the disease. From (Figure-4), the demographic based evaluation showed that majority of the pediatric patients (57.14%) belongs to hernia cases while cystic fibrosis patients were 25.71% and hypospadias patients were 17.14%. Among the hernia patients 40% were male and 17.14% female cases, among cystic fibrosis patients 17.71% were male and 8% were female cases while among hypospadias patients, 17.14% were male patients and there were no cases of female patients in hypospadias.

Table 3: Distribution of pediatric diseases

S.No.	Type of Disease	No. of Patients (%)		Total No. of Patients (%) (n=175)
		Male	Female	
1	Hypospadias	30 (17.14)	0 (0.00)	30 (17.14)
2	Hernia	70 (40.00)	30 (17.14)	100 (57.14)
3	Cystic fibrosis	31 (17.71)	14 (8.00)	45 (25.71)

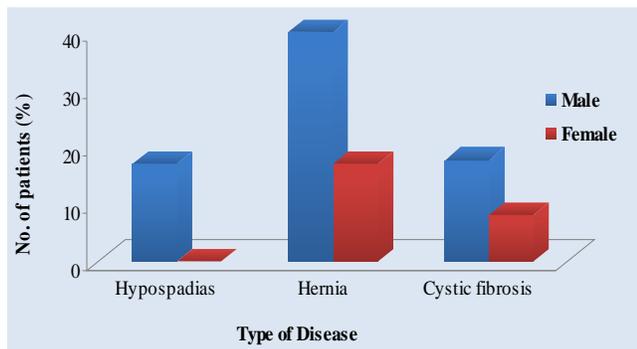


Fig 4: Distribution of pediatric diseases

Treatment involves surgery to reposition the urethral opening and its necessary, straighten the shaft of the penis. After surgery patients were given some medications like antibiotics, analgesics, anticholinergics etc. During the surgery of hypospadias, a pediatric urology surgeon uses tissue grafts from the foreskin or from the inside of mouth to reconstruct the urinary channel in the proper position, correcting the hypospadias. The following drugs are used after surgery for the recovery of the patients (Table-4):

- Antibiotics e.g. ceftriaxone, amikacin, ceftazidime etc.
- Analgesic-antipyretic drugs e.g. paracetamol, ibuprofen etc.
- Anticholinergic drug e.g. oxybutynin chloride to relax the bladder muscles and to control some itching problems which is generally occurring after surgery.
- Electrolyte fluid to recover the body glucose and electrolyte.

Although no corrective medical therapy for hypospadias is known, hormonal therapy has been used as an adjuvant for infants with exceptionally small phallic size. Preoperative treatment with testosterone injections or creams, as well as human chorionic gonadotropin (HCG) injection, has been used to promote penile growth. From (Figure-5), it was analyzed that cephalosporins were the most commonly prescribed medications after the surgery in hypospadias patients contributing to 60% cases of male patients. Aminoglycosides were prescribed in 30% cases of male patients. Anticholinergics were prescribed in 20% cases of male patients. Broad spectrum penicillin's were prescribed in 16.67% cases of male patients. Combination therapy of ampicillin with clavulanic acid was also prescribed in 16.67% cases of male patients. Electrolyte fluids and combination therapy of amikacin with ceftazidime were prescribed respectively in 13.33% and 10% cases.

Table 4: Drugs used in hypospadias

S. No	Name of Drugs	Total no. of male patients (%) (n=30)
1.	Cephalosporins	18 (60.00)
2.	Aminoglycosides	9 (30.00)
3.	Anticholinergics	6 (20.00)
4.	Broad Spectrum Penicillin's	5 (16.67)
5.	Combination therapy (Ampicillin + Clavulanic acid)	5 (16.67)
6.	Electrolyte Fluids	4 (13.33)
7.	Combination therapy (Amikacin + Ceftazidime)	3 (10.00)

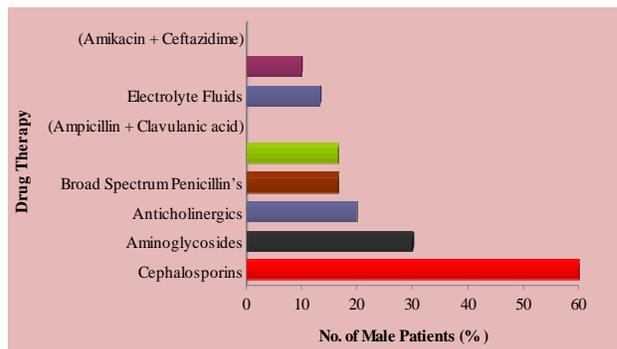


Fig 5: Drugs used in hypospadias

Drug therapy after surgical repair of hernia generally includes antibiotics, analgesic-antipyretic drugs etc. From (Table-5 and Figure-6), it was observed that combination therapy of amoxicillin with potassium clavulanate was most frequently prescribed medicine i.e. prescribed in 67% patients of hernia after the surgery. Combination therapy of diclofenac sodium with serratiopeptidase was prescribed in 41% cases of hernia, paracetamol was given in 35% cases of hernia while combination of amikacin with ceftazidime was given in 31% cases of hernia after surgery.

Table 5: Drugs used in hernia

S. No.	Name of Drug	No. of Patients (%) (n=100)
1.	Amoxicillin with Potassium clavulanate	67 (67.00)
2.	Diclofenac sodium with Serratiopeptidase	41 (41.00)
3.	Paracetamol	35 (35.00)
4.	Amikacin with Ceftazidime	31 (31.00)

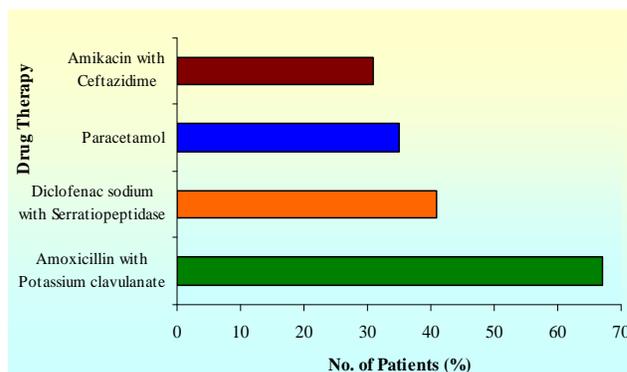


Fig 6: Drugs used in hernia

The drug therapy involved in management of cystic fibrosis cases included many drug categories as showed in (Table-6 and Figure-7). Antibiotics were the most commonly used drug category in the management of cystic fibrosis and contributed to 60% cases of cystic fibrosis. Anti-inflammatory agents contributed to 42.22% cases of cystic fibrosis. Mucolytics were used in 33.33% cases of cystic fibrosis. Bronchodilators were utilized in 26.67% cases of cystic fibrosis. Pancreatic enzyme supplements were used in 24.44% cases of cystic fibrosis while multivitamins were consumed for 17.78% cases of cystic fibrosis.

Table 6: Drugs used in cystic fibrosis

S. No.	Name of Drugs	No. of Patients (%) (n=45)
1.	Antibiotics	27 (60.00)
2.	Anti-inflammatory agents	19 (42.22)
3.	Mucolytics	15 (33.33)
4.	Bronchodilators	12 (26.67)
5.	Pancreatic enzyme supplements	11 (24.44)
6.	Multivitamins	8 (17.78)

Children's Medical Center of Dallas and University of Texas, South-western Medical Center at Dallas, USA, 2009.

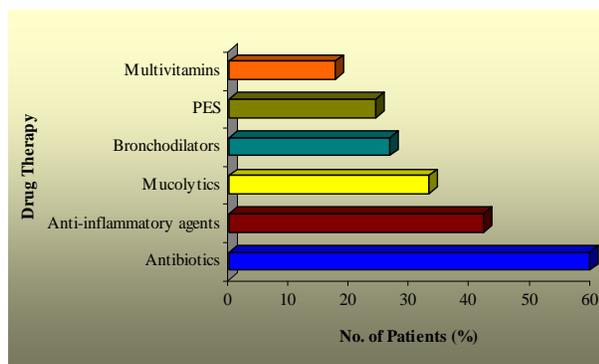


Fig 7: Drugs used in cystic fibrosis

There was no any case of adverse drug reactions and drug interactions occurred throughout the period of study as well as among the patients involved under the study.

Conclusion

It was concluded from the study that hypospadias, a disease that occurs mostly in males, is a problem of concern in the society because of its complicated pathology as well as tedious approach of treatment. Hernia and cystic fibrosis were also considered as serious problem among pediatrics but are less complicated than hypospadias. Study showed that patients of hernia were more as compared to cystic fibrosis and hypospadias which reflected that hernias are more common in patients as compared to other two diseases. So, it was suggested that being critical problems of concern, such diseases and their diagnosis, care, prevention and management need to be look after seriously in the field of medicine and educational information related to them should be discussed among the society.

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