

SMART**PEDI****UPDATE**

Dear Doctor,

Greetings from Micro Labs Limited. We are happy to bring you **"PEDI SMART NEWSLETTER"** which contain latest and interesting news in the field of Paediatrics.

This issue contains:

1. **Drug-handling problems in paediatrics and Ideal paediatric Drug Expectations by Patients and Children**
2. **Medical complications of Achondroplasia in Pediatrics**
3. **Congenital erosive and vesicular dermatosis in a Premature Neonate**

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Looking forward to your valuable feedback

Best Regards,
Medical Team, Micro Labs Limited

1. Drug-handling problems in paediatrics and Ideal paediatric Drug Expectations by Patients and Children

Introduction:

In children and adolescents, drug administration errors and drug handling issues are prevalent. In a recent review the median prevalence of 16.3 % administration errors among hospitalized pediatric patients, with errors in administration technique accounting for 53 % of these kind of errors.¹ Dosages and dosage forms suitable and recommended for children are often unavailable. Hence, drug-handling issues are especially widespread in children and adolescents. As a result, dosage formulations designed for adults must be altered for use in children like crushing and splitting of tablets. Additionally, dosage forms designed specifically for children, such as liquid formulations, involve extra processes in the preparation method compared to simple handling of tablets for adults.² Dosing, administration, the complexity of the treatment regimen, or pharmacological features such as drug formulation are instances of relevant medication-related concerns, particularly in young patients. Behavioral characteristics, such as skills in drug administration have also been found as important contributors in medication adherence.² However, it is still unknown which issues children and parents have when receiving paediatric medications. By sharing their experiences and expectations, parents and children can give useful information for the development and design of future medicine formulations for children. As a result, researchers conducted a questionnaire survey of children and parents to know about (A) their own experiences with paediatric drug administration and (B) their expectations of an ideal paediatric drug.

Expectations concerning the ideal pediatric drug was highly heterogeneous

Methods:

This is a prospective observational study (i) In- and outpatients aged 0 to 5 years, as well as (ii) patients aged 6 to 17 years and their parents, were invited to participate in a questionnaire study. A questionnaire was designed by an expert panel of pharmacists, a neuro pediatrician, and a child psychotherapist. The following key issues were addressed in the Questionnaire (Part A) Problems with drug handling Experience. (Part B) Ideal paediatric drug expectations. (B.1) Favored drug administration route. (B.2) Favored peroral drug characteristics. (B.3) Description of the ideal pediatric drug in their own words.

SMART**Pedi****UPDATE****Results:**

The study included (i) 46 parents of children aged 0–5 years and (ii) 103 paediatric patients aged 6–17 years and their parents. Experiencing drug-handling problems during drug administration was reported by 100% of parents of 46 children aged 0–5 years, 62% of 103 children aged 6–17 years, and 70% of their parents.

Of the 46 parents of children aged 0–5 years, 65% favoured a peroral route of drug administration for long-term medication, 17% preferred the rectal route, 11% preferred a dermal route, and 7% preferred an injection and others 2%.

91% of 46 parents of children aged 0–5 years said they favoured liquid drug formulations, and 7% preferred solid drug formulations. Of 103 children aged 6–17 years and their parents, 41% children and 46% parents preferred liquids; 37% children and 40% parents favoured solid dosage forms.

Preferences of 6–17-year-old children and their parents matched in 43 to 66%.

Discussion:

The main reasons of problems in drug handling by children and their parents were investigated in this study. In this study, the opinions of children and their parents on how a medicine should be administered were of specific interest. The participants in this survey also reported challenges with tablet splitting, which is supported by existing literature. The quartering of 10 mg hydrocortisone tablets was proven to create unacceptable dosage fluctuations despite functional break lines. As a result, the study's researchers advise using mini-tablets in correct dosage for children.³ In around half of the studies, children and parents had different preferences for medicine formulations. Findings of this study are in accordance with past studies that have indicated that children are certainly well able to describe experiences and expectations for their medication⁴. The responses to the open question regarding the perfect paediatric drug revealed a wide range of preferences. The responses were all quite unique, and there was no clear trend.

Conclusion:

In Conclusion, the majority of paediatric patients and their parents had already faced drug-handling issues. Children and their parents had quite different expectations about the ideal paediatric medication formulation. As a result, to improve paediatric medication therapy, individual patient expectations should be considered, and children should be contacted directly to know more information about their viewpoint, as long as they are mature and intellectually capable of expressing their expectations. Future studies should look into the association between head circumference and the needs for foramen magnum decompression, as well as body weight and orthopaedic complications.

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2. Medical complications of Achondroplasia in Pediatrics

Introduction:

The most frequent of the bone dysplasias that cause short stature is achondroplasia (dwarfism).¹ It's an autosomal dominant disorder that's usually diagnosed clinically at childbirth and then confirmed by radiological and/or genetic examinations.¹ Short stature, short limbs (proximal more than distal), macrocephaly, midfacial retrusion, small thorax, thoracolumbar kyphosis, overstated lumbar lordosis, restricted or limited elbow extension, short fingers, hip and knee laxity, and lower-leg bowing and weakness are some of the clinical characteristics of achondroplasia.² These affect children's motor development and physical health, resulting in mild to moderate physical impairment.

This information will help practitioners plan for achondroplasia management and give parents a better understanding of the likelihood and timing of complications

Medical complications such as hydrocephalus, craniosynostosis, craniocervical stenosis, subdural haematomas, ear, nose, and throat complications (hearing impairment, sleep-disordered breathing), orthopaedic complications (lower-limb malalignment, spinal stenosis, thoracolumbar kyphosis), respiratory complications, obesity, and death might even occur.² This study aimed at evaluating the rates of medical investigations, complications, interventions, and outcomes in children with achondroplasia

Methods:

It is a retrospective cohort study. The study retrospectively evaluated the medical complications in children with Achondroplasia. All the patients aged between 0 and 18 years of age, who had achondroplasia and had sought medical care at least 2 times and had confirmed radiological diagnosis or a molecular diagnosis for Achondroplasia are included in the study. The information on the date and country of birth, gender, parental achondroplasia, and date of death.

The date and method of the initial diagnosis, as well as the presence of shortened bones on prenatal ultrasound, were all documented. Data on Cranio-facial, orthopaedic, obesity, ear, nose, and throat diagnosis, complications, complication diagnosis, intervention, and outcomes following intervention were obtained and recorded from medical records. The statistical analysis was carried out using standard descriptive statistics.

Results:

A total of 116 people were identified. A total of 108 people participated in this study, with 58 men and 50 women. Ninety-nine people (91.7%) were enrolled in the research at birth. The remaining nine (8.3%) participants enrolled in the study after birth. The median exit age was 8 years and 8 months, with a median follow-up of 8 years and 8 months.

Complications	n (%)
Craniofacial complications	
Craniocervical stenosis	52 (48)
Hydrocephalus	15 (13.9)
Subdural haematomas	3 (2.8)
Craniosynostosis	1 (0.9)
Ear, nose, and throat complications	
Hearing impairment	66 (61.1)
Sleep-disordered breathing	44 (40.7)
Orthopaedic complications	
Clinically significant lower-limb malalignment	46 (42.6)
Clinically significant thoracolumbar kyphosis	24 (22.2)
Symptomatic spinal stenosis	10 (9.3)

48% of the participants had craniocervical stenosis, 13.9 % had hydrocephalus, 61.1 % had hearing loss, 40.7 % had sleep apnea, 42.6 % had lower-limb malalignment, 22.2 % had thoracolumbar kyphosis, 9.3 % had symptomatic spinal stenosis, 11.1 % had obesity, and 14.8 % had at least one admission for respiratory problems. During the research period, two deaths occurred.

Discussion:

The research provides up-to-date information about medical complications in children with achondroplasia. Unlike previous studies, all the study subjects in this study had a confirmed diagnosis of achondroplasia. The study had a mean follow-up of almost 9 years. The data in this study is complete, continuous, and retrospective. This is significant because new pharmaceutical treatments are limiting future opportunities to describe the natural history of medical issues in unmodified achondroplasia, especially in children. Craniocervical stenosis was found in 48% of those who participated in the study. When expressed as a percentage of those who underwent a head MRI, the figure was 84.3 %. This indicates that craniocervical stenosis was most likely underestimated in this study.³ The rate of preterm birth (8.3%) was similar to data suggesting that 8.7% of infants are born prematurely. 3 of 10 people with

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symptomatic spinal stenosis were obese, according to the researchers; a larger sample size is needed for detailed analysis.⁴ In contrast to other studies, this one had a higher rate of hospital admissions, with 14.8 % having at least one hospitalisation for acute respiratory symptoms. Researchers assume that the high rate of hospitalisations in this study is due to children with achondroplasia having anatomically narrow airways, a small chest due to rib shortening, and a lack of respiratory reserve.

Conclusion:

The study looked at current medical complication rates in children with achondroplasia. This information will help practitioners plan for achondroplasia management and give parents a better understanding of the likelihood and timing of complications. With the development of pharmaceutical treatments for achondroplasia, this data on the natural history of unmodified achondroplasia is very important.

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3. Congenital erosive and vesicular dermatosis in a Premature Neonate

Introduction:

Congenital erosive and vesicular dermatosis (CEVD), is an unusual disorder with unknown cause. Erythema, vesicles, erosions, crusts, and fissures typically appear at birth, affecting more than 75% of the skin. Within 10–3 months, the lesions heal on their own, leaving pathognomonic reticulate scarring. Here is a case of prematurely born boy with CEVD.^{1,2}

Congenital erosive and vesicular dermatosis should be considered in the differential diagnosis of non-infectious erythematous plaques and erosions in neonates.

Case Presentation:

The authors describe the case of a baby who was born prematurely at 25 weeks' gestation to a 31-year-old female after an emergency caesarean birth. The pregnancy had gone smoothly, with regular check-ups. The newborn was admitted to the neonatal critical care unit for severe respiratory distress syndrome (hyaline membrane illness). Although the newborn had no skin abnormalities at birth, a tiny erythematous plaque with hypopigmented regions and a cribriform surface appeared on his left flank after a few weeks. There was no obvious traumatic cause, and syphilis, hepatitis, toxoplasmosis, rubella, cytomegalovirus, and herpes simplex virus (HSV) 1 and HSV2 serological testing were negative. After 94 days, the infant had made a good recovery and was discharged from the hospital. The patient was referred to a dermatology outpatient clinic 10 months after the birth. Patient had a 3 cm cicatricial-like cribriform plaque on his left flank (figure 1), as well as a smaller identical lesion on the periumbilical region, with swollen pink tissue surrounded by a whitish perilesional halo. Scar tissue and small foci of granulomatous foreign body reaction were seen in a punch biopsy of the larger lesion. A diagnosis of CEVD was made based on the exclusion of other possible diagnoses, the course of the dermatosis, the histopathology, and the typical scarring.



Figure 1 Small erythematous plaque, first observed in the neonatal ICU



Figure 2 Scar tissue at 3 years old, note the peculiar shape and cribriform surface.

Discussion:

CEVD has been recorded in less than 40 cases. Intrauterine infections, amniotic adhesions, birth trauma, and a developmental abnormality with atypical healing in premature skin are all

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possible causes.^{1,3} Although a few occurrences have been observed in full-term infants, it is strongly connected to prematurity.² Nail abnormalities, hyperthermia/hypohidrosis, maternal chorioamnionitis, alopecia, neurodevelopmental and ophthalmological abnormalities, and tongue atrophy are some of the other associations. Postnatal herpetic superinfections may be a complication for these patients. Amniotic construction bands, congenital infections, epidermolysis bullosa, incontinentia pigmenti, focal dermal hypoplasia, aplasia cutis congenita, and autoimmune bullous diseases are all included in the differential diagnosis. The diagnosis is established after all these differentials are eliminated and the typical supple-reticulated scarring develops. ¹The histological results differ depending on the disease stage. Epidermal necrosis, subepidermal vesiculation, and an eroded epidermis with a mainly neutrophilic or mixed dermal infiltration have been reported in early inflammatory lesions, whereas scar tissue has been seen in late lesions. This patient had uncommon clinical symptoms, such as the development of lesions after birth and a limited level of skin involvement. patient is currently 3 years old (figure 2) and receives regular cicatricial tissue massages in order to maintain skin elasticity. CEVD has a good long-term prognosis, and most patients do not develop new lesions after neonatal period. The cosmetic effects of permanent scarring, however, must be considered.

Conclusion:

The study concludes that Congenital erosive and vesicular dermatosis is strongly connected to prematurity and should be considered when diagnosing non-infectious erythematous plaques and erosions in new borns.

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