Volume 4 | Issue 19

SMART PEDI UPDATE

Pedismart Newsletter
Vol 4 | Issue 19 | 2025

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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. **Identification of risk factors for late pulmonary artery hypertension in Extremely premature infants**
2. **Occurrence of avascular necrosis in the context of rheumatologic diseases**
3. **Prevalence of depression and anxiety in patients with childhood-onset systemic lupus erythematosus**
4. **Case Report on Pediatric Splenic Sclerosing angiomatoid nodular transformation (SANT)**

Should you require any specific article or medical information, please feel free to contact us at medicalservices@microlabs.in

Looking forward to your valuable feedback

Best Regards,

Medical Team, Micro Labs Limited

SMART PEDI UPDATE
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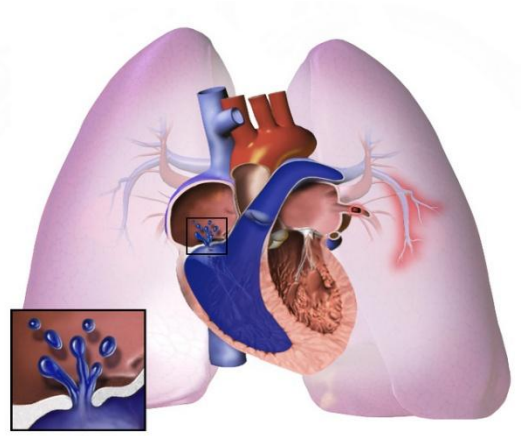
Identification of risk factors for late pulmonary artery hypertension in Extremely premature infants

Introduction



Bronchopulmonary dysplasia (BPD), a common condition affecting nearly 45% of surviving extremely preterm infants born before 28 weeks' gestational age (GA)[1], is often linked to late pulmonary artery hypertension (PH). Increased pulmonary artery pressure over ≥36 weeks postmenstrual age (PMA) is associated with higher morbidity and mortality.[2] Infants with late PH and BPD have higher readmission rates in the first year after neonatal intensive care unit discharge, as well as mortality rates of 26%-47% in the second and third years.[3] Despite the serious consequences of late PH, there is no standardized method for screening and diagnosing neonates. The timing and criteria for late PH screening in infants vary significantly. Echocardiography is commonly used to diagnose late PH, but the parameters used to assess pulmonary artery pressure vary between centers.[4] Although the pathogenesis of late PH is likely multifactorial, identifying specific risk factors remains unclear. The study aimed to address the knowledge gap by identifying risk factors and their link to the development of late PH in premature infants born before 28 weeks of gestation.

Methods

This retrospective cohort study included infants born before 28 weeks' gestation who had PH screening and echocardiography at 36 weeks' post menstrual age. Patients with major congenital anomalies or undergoing extensive cardiac surgery were excluded. Data collected included infant and maternal demographics, infant outcomes, respiratory support at 1 week, 4 weeks, and 36 weeks of PMA, patent ductus arteriosus (PDA) status and treatment, and PH screening echocardiogram results. Diagnoses, clinical findings, therapies, and echocardiogram results were collected from progress notes, electronic medical records, and echo reports. PH was determined using echo parameters such as pulmonary artery acceleration time (PAAT), ventricular septal position, and estimated systemic pressures and tricuspid valve jet regurgitation velocity. Then, the infant's demographic and clinical characteristics were compared to their late PH status to determine any correlation. The data analysis was performed using R (v.4.2.1).

Results

The study analysed 99 infants, of whom 20 (20%) developed late PH. There were no significant associations between maternal or neonatal characteristics and the development of late PH.

Procedural closure of the patent ductus arteriosus (OR 3.72, 95% CI: 1.24–11.1, p = 0.02), home oxygen use (80% vs. 58%, p = 0.04), and the FiO2% requirement at 4 weeks of age were linked to late PH. Late PH was linearly correlated with the severity of bronchopulmonary dysplasia (BPD), with a 3.5-fold increase in the odds of a late PH diagnosis for every point increase in BPD severity (Figure 1). There were no significant differences in the incidence of necrotizing enterocolitis (NEC), retinopathy of prematurity (ROP), sepsis, or intraventricular hemorrhage (IVH) between infants with and without late PH.

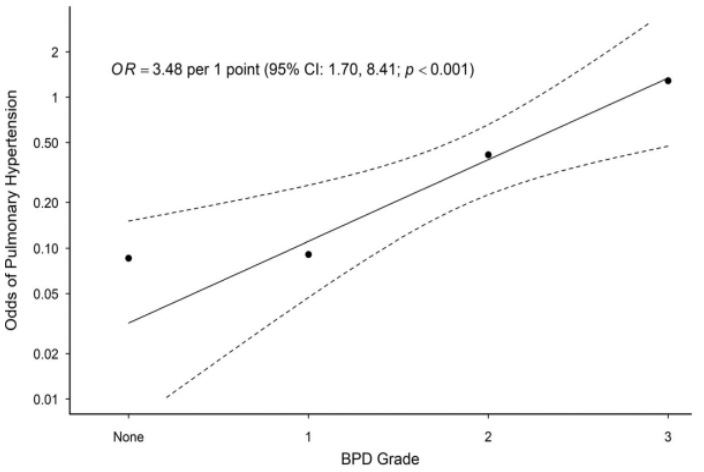


Figure 1. Relationship between odds of developing late pulmonary hypertension and BPD grade.

Discussion

This study found a strong association between BPD severity and late PH in preterm neonates born before 28 weeks' gestational age. A one-point increase in BPD grade was associated with a 3.5-fold increase in the odds of developing late PH. This suggests that impaired vascular proliferation and heterogeneous ventilation in BPD may contribute to the pathogenesis of late PH. The markers of severe respiratory disease, including FiO2 requirement at 4 weeks, postnatal steroid use, home oxygen need, and procedural PDA intervention, were linked to late PH. This is consistent with the previous study that infants with late PH were more likely to be discharged home on oxygen therapy, indicating a strong link with BPD.[5] Additionally, infants with late PH were more likely to receive procedural PDA treatment, which may indicate that PDA shunt burden plays a part in the development of late PH, as observed by Gentle et al.[6]

Conclusion

There is a strong relationship between late PH and BPD severity in extremely preterm infants, supporting the idea of a common pathophysiological mechanism. Given the increased morbidity and mortality associated with late PH, the study suggests implementing a targeted

screening program for all infants with BPD at 36 weeks' PMA to assess for late PH, followed by close monitoring after discharge.

Among infants born very prematurely, one in five experienced late PH. The onset of late PH was linked to indicators of the severity of respiratory diseases, such as the BPD grade.

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Occurrence of avascular necrosis in the context of rheumatologic diseases

Introduction

Avascular necrosis (AVN) is a severe bone and red medulla necrosis caused by traumatic events or atraumatic risk factors, such as glucocorticoid use.[1] Atraumatic AVN primarily affects long-bone epiphyses, specifically the femoral head and condyles, distal end of the tibia, and humeral head.[2] It is influenced by several genetic and metabolic factors in addition to physical blood flow-restrictive events.[3] AVN has been linked to rheumatologic disorders, particularly systemic lupus erythematosus (SLE), due to inadequate bone perfusion and the reported prevalence of AVN in SLE patients ranges from 10 to 44%.[4] There is limited evidence on AVN in children with rheumatic diseases, with only single case reports and small series available. This study investigated the occurrence of AVN in children with underlying rheumatic disease and described the demographic and clinical characteristics of AVN patients, with a focus on treatment background.

Methods

This retrospective multicentric study included patients diagnosed with AVN confirmed by MRI before the age of 18 and affected by a rheumatic disease. Exclusion criteria consists of prior diagnoses of congenital hip dysplasia, Gaucher disease, hemoglobinopathies, malignant

cancer, and/or prior hematopoietic or solid transplantation, as well as prior orthopedic surgery performed at the site of AVN. Data collection was conducted using a standard case report form (CRF) to gather information on demographics, clinical conditions, laboratory results, imaging, and treatment history, particularly steroid exposure was documented. Further assessment of the population was conducted based on the time of AVN onset and the various underlying rheumatologic diseases, i.e. systemic lupus erythematosus (SLE) and systemic juvenile idiopathic arthritis (sJIA).

Results

About 14 cases were collected from seven centers (7 with SLE, 4 with JIA, and 3 others) (Figure 1). AVN affected 71% of females, with a median age of 14.3 years at diagnosis (IQR 13.2–15.4). Most of the reported multifocal involvement (93%), primarily involving the knees (28%), and femoral heads (44%). Most patients (71%) had symptoms, with pain being the most commonly reported symptom (100%), followed by joint swelling (40%) and functional limitation (70%). All patients with severe rheumatology received systemic glucocorticoids, with a median cumulative prednisone equivalent dose of 457.5 mg/kg (IQR 259.5–849). Almost all patients (93%) experienced symptoms of steroidal toxicity. Imaging revealed persistent abnormalities in all patients except one patient, despite complete symptom resolution in 6 patients. Bisphosphonate, hyperbaric oxygen, and magnetic therapy were employed in four, three, and one case, respectively. Two patients underwent orthopedic surgery. Five patients (36%) experienced a full resolution of their symptoms along with a return to normal imaging, whereas six patients (43%), experienced permanent sequelae.

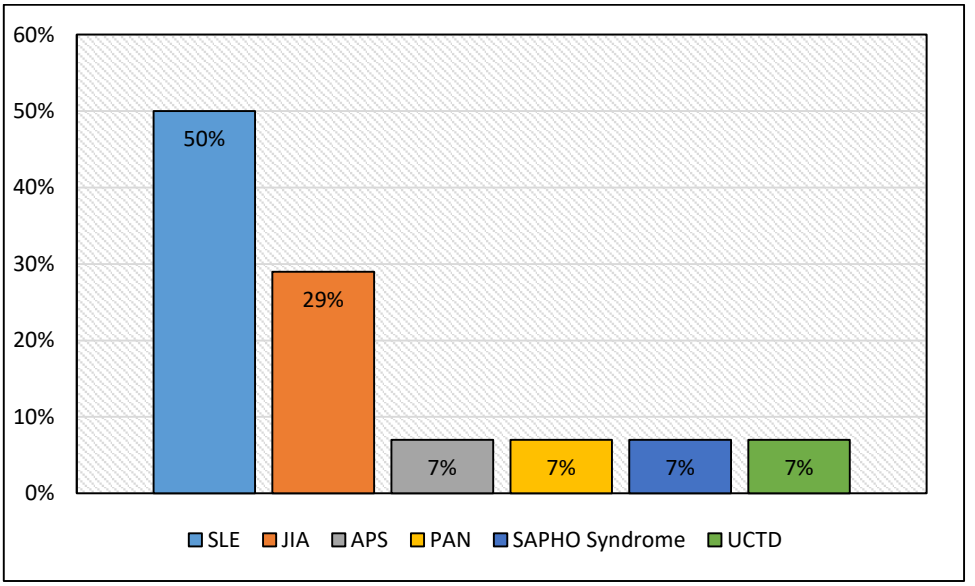

Figure 1. Rheumatologic background of collected patients with AVN

Discussion

The study identified AVN as a rare but severe complication of pediatric rheumatologic diseases, predominantly affecting patients with SLE. AVN is typically symmetrical, insidious, and asymptomatic in a significant number of cases, primarily involving the femoral head and knee. Glucocorticoids are considered as the main risk factor, with almost all patients in the cohort exhibiting clinical signs of steroid toxicity.[4] Consistent with this study, Yang et al. found that the most significant risk factor for AVN in pediatric SLE was the maximum daily prednisone dose, even though both the cumulative and maximum daily prednisone dose were significant in determining risk for AVN.[5]

Conclusion

AVN, a rare but severe complication of pediatric rheumatic diseases that must be closely and actively monitored to prevent long-term damage.

High cumulative glucocorticoid exposure is a key risk factor for avascular necrosis in pediatric rheumatic disease, highlighting the need for careful steroid dosing and proper monitoring.

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Prevalence of depression and anxiety in patients with childhood-onset systemic lupus erythematosus

Introduction

Childhood-onset systemic lupus erythematosus (cSLE) is a multi-systemic autoimmune disease that often causes depression and anxiety disorders. A few studies have been carried out and found the prevalence of clinically significant depression and anxiety in patients with cSLE, ranging from 6.7 to 59% for depressive symptoms and 34-37% for anxiety symptoms, respectively.[1] The detrimental effects of anxiety and depression that first appear in childhood and adolescence continue into early adulthood and include poor mental and physical health, substance abuse, and low educational and employment status.[2,3] Early detection of anxiety and depression disorders in children and adolescents with cSLE is therefore crucial. A previous

study found that relatives of individuals with SLE have lower health-related quality of life (HRQOL) compared to the general population, with nearly half experiencing depression and anxiety.[3] This study aimed to ascertain the prevalence of clinically significant anxiety and depression in cSLE patients, pinpoint risk factors for these symptoms, and evaluate the effects of these conditions on HRQOL and family members.

Methods

This cross-sectional study included individuals aged 8-18 years, diagnosed with cSLE using the 2012 Systemic Lupus International Collaborating Clinics (SLICC) classification criteria or the 2019 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria before the eighteenth birthday, and experiencing illness for more than three months after cSLE diagnosis to allow for adjustment. Patients with intellectual disabilities, limited Thai proficiency, or difficulty understanding or completing questionnaires were excluded from the study. Depression and anxiety were assessed using the Children's Depression Inventory (CDI) and the Screening for Child Anxiety Related Disorders (SCARED), respectively. Health-related quality of life (HRQOL) was measured using the Pediatric Quality of Life Inventory Generic Core Scale (PedsQL-GC), and pain intensity was recorded using a visual analog scale. Parents completed the Pediatric Quality of Life Family Impact Module (PedsQL family impact) to evaluate the effects on family well-being. Additional data on demographics, disease activity (SLEDAI-2K), organ damage (SLICC/ACR Damage Index), medication use, and socioeconomic background were collected. The data was analyzed using SPSS 22.0 statistical software.

Results

The study included 91 patients with cSLE, with a median disease duration of 3.4 years (IQR 3.5) and the median SLE disease activity index 2000 score was 2 (IQR 6). Clinically significant depression (CDI>15) and clinically significant anxiety (SCARED≥25) were found in 31.9% and 49.5% of people, respectively. There were 26 patients (28.6%) with coexisting clinically significant anxiety and depression. In multivariable analyses, older age at diagnosis was associated with clinically significant depression (OR 1.56, 95% CI 1.12-2.16, p=0.008). Organ damage (OR 4.27, 95% CI: 1.19-15.31, p=0.026) and pain score (OR 1.61, 95% CI: 1.11-2.32, p=0.012) were associated with clinically significant anxiety. Patients experiencing clinically significant depression or anxiety had significantly lower PedsQL-GC and PedsQL family impact scores than those without these symptoms (Figure 1).

Variables	CDI	SCARED	Pain Score	SCLEDOI-2K	SDI	PedsQL-GC	PedsQL family impact
CDI	1.00						
SCARED	0.71**	1.00					
Pain Score	0.44**	0.41**	1.00				
SCLEDOI-2K	0.22*	0.21*	0.26*	1.00			
SDI	0.12	0.27*	0.35**	0.25*	1.00		
PedsQL-GC	-0.65**	-0.76**	-0.41**	-0.22*	-0.25*	1.00	
PedsQL family impact	-0.38**	-0.35**	-0.23*	-0.19	-0.25*	0.48**	1.00

*p-value<0.05, **p-value<0.01

Figure 1. Correlations between depressive, anxiety, pain, disease activity, damage index, HRQOL, and family impact scores

Discussion

The study found that 31.9% of patients with cSLE had clinically significant depression and 49.5% had clinically significant anxiety. Of these patients, 28.6% had both clinically significant depression and anxiety. Clinically significant depression was associated with older age at diagnosis, while anxiety was linked to organ damage and pain score. Patients with clinically significant depression and/or anxiety had lower HRQOL scores across all domains compared to those without such symptoms.[3] In line with this study findings, a previous study has shown that children with cSLE have a lower HRQOL than children in good health. Anxiety and depression symptoms were found to be associated with a lower HRQOL score among other factors.[4]

Conclusion

These findings indicate that depression and anxiety are prevalent in cSLE and negatively impact HRQOL and family life. Physicians should be aware of these psychological symptoms, especially in patients with risk factors. Providing psychological counseling and timely referrals to psychiatrists can improve HRQOL and family functioning.

Routine screening for depression and anxiety, combined with effective pain and organ damage management, is essential for optimizing quality of life in children with SLE.

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Case Report on Pediatric Splenic Sclerosing angiomatoid nodular transformation (SANT)

Introduction

Lymphomas are among the most common diseases that can affect the spleen. However, non-lymphoid tumors of the spleen are rare and primarily vascular in origin. Sclerosing angiomatoid

nodular transformation (SANT) is a rare vascular non-neoplastic lesion that has an unclear etiology and a benign clinical course. The majority of reported cases of splenic SANTs are in adults, particularly middle-aged women. Previous study indicates that SANTs have multiple angiomatoid nodules separated by dense fibrous stroma as their histological characteristics and these features are closely associated with the radiological appearance and spoke-wheel pattern.[1] Currently, there have only been 10 cases reported to date in pediatric patients (aged <18 years). This case report examines the radiological and clinical characteristics of a case of SANT in an 8-year-old boy.

Case Presentation

A 2.5-year-old child presented with new onset ptosis of the left eye and headache. The child's parents denied having any history of trauma or insect bites. During the physical examination, the child showed ptosis, exotropia, and mydriasis in the left eye. An ophthalmological examination revealed an exotropically deviated and abducted left eye, with a relative afferent pupillary defect and no signs of nystagmus. Upon slit lamp examination, the anterior segment was found to be unremarkable, with no evidence of abnormal pathologic findings. Unfortunately, we were unable to assess visual acuity due to the child's refusal to cooperate. The neurological examination revealed otherwise intact functional cranial nerves, with no evidence of cerebellar signs. Upon admission to our ophthalmology department, the child underwent a comprehensive evaluation, including a contrast-enhanced head CT, lumbar puncture, abdominal ultrasound, and immunological and viral panels. All results were normal.The CTA revealed no aneurysms in the anterior or posterior communicating arteries. A standard MRI was then performed, but this examination, too, revealed no lesions. During hospitalization, a high-resolution MRI was performed with 0.5 to 1 mm slice thickness. This advanced imaging technique successfully identified a 5 mm diameter ovoid mass in the third cranial nerve, superior to the internal carotid artery (ICA). 3T MRI scanner was used, resulting in higher signal strength and better visualization of lesions compared to traditional MRI. The lesion showed an isointense signal on T1-weighted images, which provided additional details. A high-resolution 3D T2-weighted MRI with a 0.5 mm slice thickness revealed a hyperintense and homogeneous lesion. An orbital MRI was also requested but found no anomalies. Prednisolone was administered at 15 mg twice daily for seven days, then tapered down to 3 mg for an additional week. On discharge, the child showed clinical improvement, including the ability to open his left eye and slightly improved ocular movements. The child underwent Botox injections to the lateral rectus muscle at another medical center due to lesions compressing the oculomotor nerve.

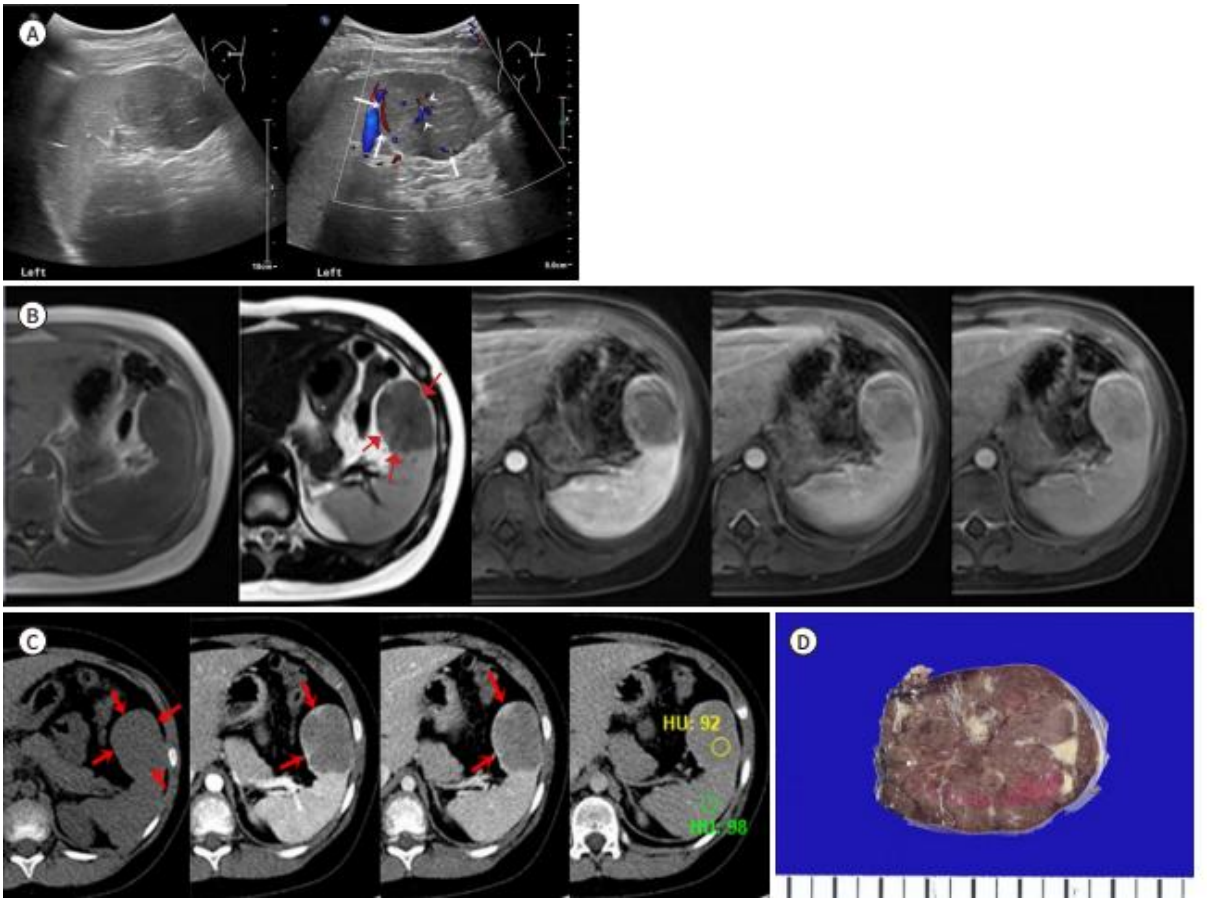

Figure 1. Splenic sclerosing angiomatoid nodular transformation in an 8-year-old patient.

Discussion

The study concludes that isolated oculomotor schwannoma in pediatric populaton is a very rare condition. When clinical symptoms like third nerve palsy or increased intracranial pressure are suspected, a thorough diagnostic approach is required to rule out other congenital or acquired conditions. High resolution MRI is essential for detecting small lesions, which may be missed with standard imaging. Steroid therapy improved the patient's symptoms, and botulinum toxin injections helped manage ocular misalignment, supporting a conservative approach over immediate surgical intervention.[3] In line with our findings, Scott AB et al demonstrated that Botulinum toxin injection is an effective adjunct therapy, preventing the contracture of the antagonist lateral rectus muscle, facilitating improved ocular alignment, and enhancing visual function.[4]

Conclusion

SANT in the spleen manifests as a well-defined mass with or without internal calcification. The mass demonstrates peripheral vascularity on US and progressive peripheral enhancement on CT and MRI. A spoke-wheel pattern on T2-weighted and contrast-enhanced imaging may be indicative of central fibrosis. Lymphoma or other benign splenic lesions like hemangioma,

hamartoma, or IPT are important differential diagnoses. When pediatric patients display these characteristics, radiologists should be aware of the radiological features of SANT and its differential diagnosis.

Pediatric splenic SANT is rare and exhibits a distinctive spoke-wheel pattern on imaging that differentiates it from malignant lesions.

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