centros; volume coletado, os quimioterápicos e o corticóide utilizados variaram entre os centros e foram de acordo com protocolo de tratamento utilizado. Da amostra/análise: o tempo de processamento de amostras foi de até 1 hora em 12/20 centros respondedores e entre 2-3h em 3 centros (utilização de conservante); 85,7% dos centros fazem análise citomorfológica independente da contagem de células sendo 52,4% realizada por bioquímico; 11/21 tem avaliação liquorica por ICF disponível, e na maioria dos centros é realizada ao diagnóstico ou na suspeita de recaída. O guia de recomendações gerado a seguir contemplou 4 partes: I- Punção lombar e coleta de LCR, II- Quimioterapia intratecal, III- Processamento do LCR, IV- Análise Imunofenotípica de LCR. Conclusão: O questionário permitiu identificar uma variabilidade nas etapas de coleta, manuseio de quimioterapia intratecal e análise de LCR no cenário brasileiro, com possibilidade de impacto direto no sucesso terapêutico dos pacientes pediátricos. Esses achados justificaram a criação de um guia de boas práticas em punção e análise de LCR, baseado em literatura e experiência dos centros participantes, com o objetivo de equalizar a rotina do oncologista/hematologista pediátrico, obter reprodutibilidade dos processos e o benefício final dos pacientes pediátricos.

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PEDIATRIC CEREBRAL SINUS VENOUS THROMBOSIS (CSVT) IN A BRAZILIAN SINGLE-CENTER: A 3-YEAR RETROSPECTIVE ANALYSIS OF A MULTIDISCIPLINARY EXPERIENCE

AA Antunes, MB Swain, DDC Bispo, RBR Pereira, BO Lima, MF Marcelino, IMQS Magalhães

Hospital da Criança de Brasília José de Alencar, Brasília, DF, Brazil

Objectives: CSVT is rare in pediatric patients but is related to considerable morbidity and mortality. Its etiology is multifactorial: infections, head trauma, cancer and some chronic inflamatory diseases are some of the risk factors. Clinical manifestations are variable and non specific: headache, seizure, focal neurologic deficit and impaired consciouness. Pathogenesis involves venous obstructive ouflow which results in impaired drainage of cerebrospinal fluid and increased intracranial pressure. Diagnosis involves a high suspicious index and an adequade neuroimaging. Management consists of suportive care, anticoagulation and treatment of the neurologic complications. This sudy has the purpose of describing a cohort of pediatric CSVT. **Materials and methods:** Patients under 18 years diagnosed with CSVT between august 2021 to august 2024 were retrospectively included. Demographic informations, signs and symptoms, laboratory parameters, management information, venous recanalization rate, chronic complications and follow-up were evaluated. **Results:** 22 patients were included, 72.7% male and 27.2% female. Median age at diagnosis was 9.8 years (0.16–17). Headache was the most common clinical manifestation (72.2%). In 90.9% of patients at least one risk factor for CSVT was present and infections were the most common (40.9%). 63.6% had more than one sinus affected and 22.7% had cerebral venous infarct. Median values of aPTT, PT, fibrinogen and platelet count were normal and median D-dimer was 3.598ng/mL (275-21373). Complete treatment and follow-up data were available in 17 patiens. Treatment included anticoagulation with enoxaparin in all patients and 2 completed antithrombotic therapy with rivaroxaban. Acetazolamide and lumbar puncture were necessary in 35.2% for high intracranial pressure control. One patient (5%) needed surgical thrombectomy. Median time of anticoagulation was 5.4 months (2.6-13.5) with 62.5% of patients with complete venous recanalization. There was no death related to CSVT but 11.7% had chonic motor sequelae. Median time of follow-up was 1.4 years (0.3-2.3) with 5.8% of recurrence thrombosis. **Discussion:** This is the first brazilian cohort of pediatric CVST. Infections were the most common risk factor. There were no clear correlation between number of sinuses affected and acute and chronic neurologic complications. Despite a small number of patients, some patients were treated with direct oral anticoagulants. The cooperation of hematology, neuroradiology, neurology and neurosurgery was essential. This cohort has limitations and some similar findings of other studies, but it highlights the role of pediatric hematologists in the approach of thromboembolic events, and also emphasizes the need of multidisciplinarity from diagnosis to acute and chronic management of pediatric thrombosis. **Conclusion:** CVST as other pediatric thrombosis needs a multiciplinary team for adequate acute and long term treatment. Multicentric studies are needed to establish more concrete data about CVST in pediatric population in Brazil.

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INTERACTION BETWEEN HAPLOTYPES OF BCL11A AND HBS1L-MYB, MAJOR QUANTITATIVE TRAIT LOCI FOR FETAL HEMOGLOBIN (HBF) LEVELS, AND INCIDENCE OF CLINICAL PHENOTYPES IN CHILDREN WITH SICKLE CELL ANEMIA

NF Sillva [a], LN Oliveira [a], AR Belisário [b], MB Viana [c], MR Luizon [a], RR Sales [a]

[a] *Universidade Federal de Minas Gerais (UFMG), Belo Horizonte, MG, Brazil*
[b] *Centro de Tecidos Biológicos de Minas Gerais, Fundação Hemominas, Belo Horizonte, MG, Brazil*
[c] *Faculdade de Medicina/NUPAD, Universidade Federal de Minas Gerais (UFMG), Belo Horizonte, MG, Brazil*

Sickle cell anemia (SCA) is a monogenic disease characterized by HbSS genotype with a complex pathophysiology. Fetal hemoglobin(HbF) is the major modifier of clinical phenotypes, and increased HbF levels inhibit polymerization of HbS and ameliorate clinical severity of SCA. GWAS have identified the BCL11A gene and HBS1L-MYB intergenic region as major quantitative trait loci for regulation of HbF expression. We identified functional SNPs of BCL11A and HBS1L-MYB