carcinoma medular. A hematúria constitui apresentação clínica comum e, geralmente, autolimitada na Anemia falciforme, não obstante, no Traço Falciforme é pouco documentada a ocorrência de hematúria com repercussão sistêmica e evolução com anemia sintomática.

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NEWBORN SCREENING FOR SICKLE CELL DISEASE IN RIO GRANDE DO SUL: EIGHT-YEAR ANALYSIS OF THE STATE REFERENCE SERVICE

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Introduction: Sickle cell disease (SCD) is a group of inherited hemoglobinopathies with autosomal recessive inheritance, characterized by the presence of the Hb S allele in homozygosity (sickle cell anemia – SCA, Hb SS phenotype) or in association with other variants (Hb C, Hb D, Hb E, β-thalassemia, among others). The Hb S mutation results from the substitution of glutamic acid by valine at position 6 of the β-globin chain (β6 Glu→Val), leading to polymerization of deoxygenated hemoglobin, erythrocyte deformity, hemolysis, and vaso-occlusive events. Symptoms usually appear after the first six months of life, making newborn screening a key strategy. In Brazil, SCD was incorporated into the National Newborn Screening Program (PNTN) in 2001, enabling early diagnosis, treatment, and lifelong multidisciplinary follow-up. **Objectives:** To evaluate eight years of newborn screening for SCD at the State Reference Service in Rio Grande do Sul (SRTN/RS). **Material e methods:** Cross-sectional, quantitative, population-based study from January 1, 2017, to December 31, 2024. All newborns screened at the SRTN/RS were analyzed for hemoglobin profile using high-performance liquid chromatography (HPLC) and/or isoelectric focusing (IEF). For those diagnosed with SCD, gender, race/skin color as reported by the parent or guardian, and age at first consultation were recorded. **Results:** A total of 776,564 newborns were screened, of which 111 (0.014%) presented an SCD phenotype. The most prevalent phenotype was SS (n = 83; 74.77%), followed by SC (n = 21; 18.92%), CC (n = 5; 4.50%), and DD, and DS (n = 1 each; total 1.80%). The overall incidence was 1:6,934 screened newborns. Among affected newborns, 63 (56.75%) were male and 48 (43.24%) female. Regarding race/skin color, 48 (43.24%) were Black, 38 (34.23%) White, 15 (13.51%) Brown (mixed race), and 11 (9.90%) other categories. The mean age at first consultation was 32 days. **Discussion and conclusion:** SS and SC phenotypes were the most frequent, with a slight predominance in males. The mean referral time (32 days) met the PNTN target (< 45 days), allowing for early antimicrobial prophylaxis and specialized follow-up. Despite the state's diverse ethnic composition, a higher prevalence was observed among those reported as Black, consistent with the African origin of the Hb S mutation. The incidence in Rio Grande do Sul is lower than in Northeastern states such as Bahia (1:650 live births), reflecting differences in Brazilian population ancestry. Newborn screening in Rio Grande do Sul enables early diagnosis of SCD and the timely initiation of follow-up. However, the current coverage (75.75% of live births within the public health network) limits the accuracy of prevalence estimates. Expanding coverage to include data from the private healthcare sector is essential to encompass all socioeconomic groups and to enhance the overall effectiveness of the program.

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O USO DE CROVALIMABE NO TRATAMENTO DA HEMOGLOBINÚRIA PAROXÍSTICA NOTURNA

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Introdução: A Hemoglobinúria Paroxística Noturna (HPN) é uma doença hematológica rara, causada por uma mutação no gene PIGA, que resulta em deficiência na biossíntese de glicosilfosfatidilinositol (GPI). Essa deficiência leva à ausência das proteínas CD55 e CD59 nas hemácias, aumentando a suscetibilidade à hemólise intravascular intensa. Entre as opções convencionais de tratamento para a HPN, destaca-se a administração de anticorpos monoclonais, cuja principal ação é a inibição do sistema complemento, sendo os medicamentos mais utilizados o eculizumabe e o ravulizumabe. Recentemente, o Crovalimabe, um novo anticorpo monoclonal humano, foi autorizado para uso no Brasil, tendo um mecanismo de ação e administração diferenciada dos medicamentos supracitados, o que gera benefícios para o paciente.