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1- Hemoglobin, 44(1):1-9, 2020. https://doi.org/10.1080/03630269.2020.1731530

Hb S/β-Thalassemia in the REDS-III Brazil Sickle Cell Disease Cohort: Clinical, Laboratory and Molecular Characteristics

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We described the clinical, laboratory and molecular characteristics of individuals with Hb S (HBB: c.20A>T)/β-thalassemia (Hb S/β-thal) participating in the Recipient Epidemiology and Donor Evaluation Study (REDS-III) Brazil Sickle Cell Disease cohort. HBB gene sequencing was performed to genotype each β-thal mutation. Patients were classified as Hb S/β0-thal, Hb S/β+-thal-severe or Hb S/β+-thal based on prior literature and databases of hemoglobin (Hb) variants. Characteristics of patients with each β-thal mutation were described and the clinical profile of patients grouped into Hb S/β0-thal, Hb S/β+-thal and Hb S/β+-thal-severe were compared. Of the 2793 patients enrolled, 84 (3.0%) had Hb S/β0-thal and 83 (3.0%) had Hb S/β+-thal; 40/83 (48.2%) patients with Hb S/β+-thal had mutations defined as severe. We identified 19 different β-thal mutations, eight Hb S/β0-thal, three Hb S/β+-thal-severe and eight Hb S/β+-thal. The most frequent β0 and β+ mutations were codon 39 (HBB: c.118C>T) and IVS-I-6 (T>C) (HBB: c.92+6T>C), respectively. Individuals with Hb S/β0-thal had a similar clinical and laboratory phenotype when compared to those with Hb S/β+-thal-severe. Individuals with Hb S/β+-thal-severe had significantly lower total Hb and Hb A levels and higher Hb S, white blood cell (WBC) count, platelets and hemolysis markers when compared to those with Hb S/β+-thal. Likewise, individuals with Hb S/β+-thal-severe showed a significantly higher occurrence of hospitalizations, vaso-occlusive events (VOE), acute chest syndrome (ACS), splenic sequestration, blood utilization, and hydroxyurea (HU) therapy.

2- Haemophilia, 26(3): e130-e133, 2020. https://doi.org/doi:10.1111/hae.13944

Hepatitis C and history of FVIII inhibitor development in a long-term cohort of Brazilian patients with haemophilia A

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Letter. No abstract available

3- Transfusion Medicine, 30(2): 148-156, 2020. https://doi.org/10.1111/tme.12653

The screening of rare blood donors in a highly admixed population: A new approach for Holley and Diego genotyping and impact of genomic and self-reported ancestry

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Objectives: The present study aimed to develop strategies for genotyping DO*HY (Dombrock system) and DI*A/DI*B (Diego system) alleles and to evaluate the impact of genomic and self-declared ancestry on rare donor screening in admixed populations.

Background: The antigens Hy and Dib demonstrate clinical importance. The lack of antisera for the serological evaluation of these antigens makes it necessary to develop molecular methods. In addition, considering that some rare red blood cell phenotypes present differences in frequency between ethnic groups, it is important to assess the applicability of self-declared ancestry in the search for rare donors in admixed populations.

Methods: DO*HY and DI*A/DI*B genotyping based on real-time polymerase chain reaction (PCR) was standardised. A total of 457 blood donors clustered by self-defined skin colour/race categories were genotyped. Furthermore, individual genomic ancestry was used in the analyses.

Results: The assays developed are reproducible and provide satisfactory results even at low concentrations of DNA, which make them useful in situations where the DNA is scarce, such as dried blood spots on filter paper, or when screening for pooled samples. No significant difference was observed in the frequencies of the DI*A, DI*B and DO*HY, comparing the self-declared White (branco) donors with those who are Black (preto) and Brown (pardo).

Conclusion: Real-time PCR, especially using pooled samples, is a promising strategy to screen rare blood donors. Although both self-reported race/colour and some blood group phenotypes are associated with ancestry, the results point to a greater complexity in the application of self-declared race/colour in the screening of rare donors in admixed populations.

4- Blood Cells, Molecules & Diseases, 80: 102376, 2020.
https://doi.org/10.1016/j.bcmd.2019.102376

Polymorphisms in genes that affect the variation of lipid levels in a Brazilian pediatric population with sickle cell disease: rs662799 APOA5 and rs964184 ZPR1

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This cross-sectional study investigated associations between SNPs in metabolizing lipid genes, alpha-thalassemia and laboratory parameters in two forms of sickle cell disease (SCD), sickle cell anemia (SCA) and hemoglobin SC disease (HbSC) in a pediatric population. Among the groups SCA and HbSC was found a higher proportion of increased triglycerides (TG) in SCA. High levels of TG were significantly associated with lower hemoglobin ($p = 0.006$) and HDL-C ($p = 0.037$), higher white blood cell count ($p = 0.027$), LDH ($p = 0.004$) and bilirubins ($p < 0.05$) in SCD. Patients with HDL-C ≤40 mg/dL had higher markers hemolytic levels. Therapy of HU significantly influenced several hematological and biochemical parameters but not lipid fractions. Genotypes of the APOA5 rs662799 were not associated with lipid levels. The G-risk allele rs964184/ZPRI ZNF259/ZPR1 gene (GC + GG genotypes) was associated with increased levels of TG in children ≥10 years old ($p = 0.045$) and the atherogenic ratio TG/HDL-C ($p = 0.032$) in SCD. The use of HU improves levels of hemolysis and inflammation markers in SCD with high TG and, while not interfering with lipid levels, seems to overlap the effect of the G-risk allele in on them. This study reported for the first time that rs964184 SNP could be a genetic modifier of TG in SCD.

5- Journal of Infusion Nursing, 43(1): 24-32, 2020.
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Peripheral Venipuncture Education Strategies for Nursing Students: An Integrative Literature Review

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This integrative literature review identified strategies to teach peripheral venipuncture to nursing students. The following databases were searched for primary studies: Biblioteca Virtual em Saúde (BVS), PubMed, Web of Science, Education Resources Information Center (ERIC), SCOPUS, and Cumulative Index to Nursing and Allied Health Literature (CINAHL). The final sample was composed of 24 studies. The literature ranged from descriptive studies to controlled clinical trials and methodologic studies to construct products/instruments for teaching peripheral venipuncture. The most frequently identified teaching strategies were theoretical contents taught via theoretical lecture, e-learning courses, video lessons, and demonstration by specialists combined with practical exercises using a mannequin, human arms, and/or haptic devices. Despite the different methods used currently, the best patient outcomes were achieved when the student received the theoretical content in an educational setting before the practical training on a mannequin and/or a virtual simulator.

6- Cytokine, 125: 154800, 2020. https://doi.org/10.1016/j.cyto.2019.154800

Evidence for interactions between inflammatory markers and renin-angiotensin system molecules in the occurrence of albuminuria in children with sickle cell anemia

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Sickle cell anemia (SCA) is an important cause of chronic kidney disease, but its pathophysiology is not completely understood. The aim of this study was to compare inflammatory biomarkers in urine samples of SCA children with and without albuminuria, and to explore correlations with renin-angiotensin system (RAS) molecules. A cross-sectional study of 213 children selected from the Minas Gerais state cohort were assigned to two groups: Group 1-89 children with SCA who had albuminuria; Group 2-124 children with SCA and normal albuminuria matched by age and sex with group 1. A subset of 89 children was prospectively followed for a median time of 1.1 year. Inflammatory biomarkers (chemokines and cytokines) in urine were measured using cytometric beads array, and RAS molecules were measured by ELISA. Children with albuminuria had significantly higher urinary levels of IP-10/CXCL10, MCP-1/CCL2, MIG/CXCL9, IL-8/CXCL8, IL-12p70, TNF, IL-10, and IL-6 than patients with normal albuminuria. In the correlation analysis, albumin/creatinine ratio was significantly and positively correlated with IP-10/CXCL10, MCP-1/CCL2, MIG/CXCL9, IL-8/CXCL8, TNF, IL-10, and IL-6. Significant correlations were found between inflammatory and RAS molecules. In the prospective analysis, cumulative risk of persistent albuminuria was higher for children with urinary levels of IP-10/CXCL10 or IL-6 above the 50th percentile. Our data showed that inflammatory markers and RAS molecules might contribute to the occurrence of albuminuria in children with SCA, suggesting that both pathways interact in sickle cell nephropathy.

7- Research and Practice in Thrombosis and Haemostasis, 4(5): 752-760, 2020.
https://doi.org/10.1002/rth2.12335

Development of inhibitors in hemophilia A: An illustrated review

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This illustrated review focuses on the development of inhibitors in patients with congenital hemophilia, which is the most serious treatment-related complication in these patients. Hemophilia A (HA) is an inherited X-linked bleeding disorder affecting 1:5000-10 000 newborn males worldwide. It results from the deficiency of coagulation factor VIII (FVIII), due to mutation(s) in its coding gene (F8). Treatment requires administration of FVIII-containing products either on demand or as prophylaxis, which can induce inhibitor development in 20%-35% of patients. Inhibitors are alloantibodies that neutralize the procoagulant activity of exogenous FVIII. During the initial administration of FVIII-containing products, patients with HA can develop a proinflammatory immune response with synthesis of anti-FVIII IgG1, which has no FVIII inhibitory activity. However, in patients with inhibitors, immune response shifts toward an anti-inflammatory/regulatory pattern favoring the synthesis of anti- FVIII IgG4 antibodies. Patients with inhibitors present with bleeding episodes that are difficult to control, and they have reduced response to FVIII replacement. Currently, immune tolerance induction is the available treatment for eradication of persistent high-

titer inhibitors. Despite the clinical relevance, the immunological mechanisms for inhibitor development in patients with HA remains unexplained.

8- Transfusion, 60(8): 1713-1722, 2020. https://doi.org/10.1111/trf.15818

Blood utilization and characteristics of patients treated with chronic transfusion therapy in a large cohort of Brazilian patients with sickle cell disease

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Background: Red blood cell (RBC) transfusions are used in sickle cell disease (SCD) to treat acute complications or as chronic transfusion therapy (CTT) to prevent severe manifestations. The objectives of this study were to describe blood utilization and adverse events (AEs) associated with RBCs in the Brazilian SCD population and compare characteristics of patients treated or not with CTT.

Study design and methods: A SCD cohort was established at six Brazilian centers. Medical and blood bank records were abstracted for clinical and transfusion history. Two controls not treated with CTT matched on center, SCD genotype, sex, and age were selected for each CTT case within the cohort to compare characteristics between the two groups.

Results: Most of the 2794-member cohort had received a transfusion (75.0% of children and 89.2% of adults) with 29.2% of patients receiving transfusion in the prior year. There were 170 (10.6%) children and 115 (9.2%) adults treated with CTT. Children not treated with CTT were more likely to have pain and acute chest hospitalizations in the prior year (25.3% vs. 11.9%, p = 0.0003; and 22.0% vs. 10.7%, p = 0.002, respectively). Both iron overload and alloimmunization were more common in CTT cases compared to controls (65.6% vs. 17.0% and 36.2% vs. 15.9%, respectively). A higher proportion of adults treated with CTT demonstrated oxygen saturation of greater than 95% compared to controls not treated (51.1% vs. 39.2%), while there was no difference in oxygenation between children treated or not. Of 4501 transfusion episodes, 28 (0.62%) AEs were reported. There was no difference in AEs associated with transfusions for acute indications versus CTT.

Conclusion: Red blood cell transfusion was common in Brazilian SCD patients, with utilization driven by CTT. Transfusion reactions were not common; however, alloimmunization and iron overload were frequent among those on CTT, highlighting the need for novel clinical strategies to mitigate these risks.

9- American Journal of Hematology, 95(5): E125-E128, 2020. https://doi.org/10.1002/ajh.25763

Prevalence and risk factors for albuminuria and glomerular hyperfiltration in a large cohort of children with sickle cell anemia

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No abstract available

10- The International Journal of Cardiovascular Imaging, 36(11): 2145-2153, 2020.
https://doi.org/10.1007/s10554-020-01924-3

Prognostic value of left ventricular longitudinal strain by speckle-tracking echocardiography in patients with sickle cell disease

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Cardiovascular complications have been increasingly detected in patients with sickle cell disease (SCD). Two-dimensional speckle-tracking echocardiography (STE) detects early myocardial changes in a number of pathophysiological processes, which may be useful in SCD. This study was designed to examine the value of STE in predicting clinical outcome in adult patients with SCD. A total of 219 patients, mean age 33 ± 12 years were prospectively enrolled. Several clinical, laboratory and echocardiographic variables including left ventricular global longitudinal strain (LVGLS) by STE were assessed. The endpoint was a composite of the following events: (1) all-cause mortality, (2) three or more acute painful episodes that require hospitalization in one year, (3) acute chest syndrome and (4) hospitalization due to disease complication. The majority of the patients had enlargement of LV and left atrial (LA) with preserved ejection fraction. During the mean follow-up of 30 months, 69 patients (32%) had reached the endpoint, including eight deaths (3.7%). No difference was observed in the parameters of diastolic function comparing the patients with and without events. LVGLS ranged from - 12.25 to - 25.44 (mean - 20.26 ± 2.5), with higher values in the patients who had events compared with those who did not. In the multivariable analysis, higher LVGLS values were associated with adverse events (adjusted OR 1.25; 95% CI 1.04-1.51; p = 0.021), independently of the TR maximal velocity and LV ejection fraction. In patients with SCD, higher LV global longitudinal strain was a predictor of adverse outcome, independently of age, TR velocity and LV function.

11- Revista da Sociedade Brasileira de Medicina Tropical, 53: e20190491, 2020.
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Hemophagocytic lymphohistiocytosis secondary to visceral leishmaniasis in an endemic area in the north of Minas Gerais, Brazil

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Introduction: Visceral leishmaniasis (VL) is an ill-studied disease that is endemic to several regions of Brazil. It is often complicated by hemophagocytic lymphohistiocytosis (HLH), a potentially fatal disorder resulting from excessive non-malignant activation/proliferation of T lymphocytes and macrophages. Considering the overlapping clinical and laboratory characteristics of these diseases, diagnosing HLH is a challenge. Therefore, tracking the association between VL and HLH is necessary in endemic areas. Although HLH can be inapparent and resolve with antileishmanicides, this may not always occur. HLH causes high lethality; therefore, immunosuppressive therapy should be instituted immediately in order to avoid a fatal outcome.

Methods: We described the epidemiological, clinical, laboratory, and therapeutic profile of this association in a region of Brazil endemic for VL.

Results: We presented 39 patients with this association in a retrospective cohort of 258 children who were admitted from January 2012 to June 2017. Of the 39 patients, 31 were from urban areas (79.5%), and 21 (53%) were males. The mean age and weight were 2.86 (2.08) years and 14.03 (5.96) kg, respectively. The main symptoms were fever (100%), hepatosplenomegaly (100%), pallor of the skin and mucosa (82.5%), edema (38.5%), bleeding (25%), and jaundice (7.5%). Hemophagocytosis was identified in 16/37 (43.24%) patients, and direct examination revealed that 26/37 (70.27%) patients were positive for VL. The patients were treated as recommended by the Ministry of Health.

Conclusions: It was observed that HLH is a common complication in endemic areas, and its diagnosis must consider the overlapping of clinical characteristics and pancytopenia.

12- Annals of Hematology, 99(7): 1453-1463, 2020. https://doi.org/10.1007/s00277-020-04079-2

Functional polymorphisms of BCL11A and HBS1L-MYB genes affect both fetal hemoglobin level and clinical outcomes in a cohort of children with sickle cell anemia

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Fetal hemoglobin (HbF) ameliorates clinical severity of sickle cell anemia (SCA). The major loci regulating HbF levels are HBB cluster, BCL11A, and HMIP-2 (HBS1L-MYB). However, the impact of noncoding single-nucleotide polymorphisms (SNPs) in these loci on clinical outcomes and their functional role on regulating HbF levels should be better elucidated. Therefore, we performed comprehensive association analyses of 14 noncoding SNPs in five loci with HbF levels and with clinical outcomes in a cohort of 250 children with SCA from Southeastern Brazil, and further performed functional annotation of these SNPs. We found SNPs independently associated with HbF levels: rs4671393 in BCL11A (β-coefficient = 0.28), rs9399137 in HMIP-2A (β-coefficient = 0.16), and rs4895441 in HMIP-2B (β-coefficient = 0.15). Patients carrying minor (HbF-boosting) alleles for rs1427407, rs93979137, rs4895441, rs9402686, and rs9494145 showed reduced count of reticulocytes ($p < 0.01$), while those carrying the T allele of rs9494145 showed lower white blood

cell count (p = 0.002). Carriers of the minor allele for rs9402686 showed higher peripheral saturation of oxygen (p = 0.002). Patients carrying minor alleles in BCL11A showed lower risk of transfusion incidence rate ratio (IRR ≥ 1.3; p < 0.0001). This effect was independent of HbF effect (p = 0.005). Carriers of minor alleles for rs9399137 and rs9402686 showed lower risk of acute chest syndrome (IRR > 1.3; p ≤ 0.01). Carriers of the reference allele for rs4671393 showed lower risk of infections (IRR = 1.16; p = 0.01). In conclusion, patients carrying HbF-boosting alleles of BCL11A and HMIP-2 were associated with milder clinical phenotypes. Higher HbF concentration may underlie this effect.

13- Transfusion, 60(2): 343-350, 2020. https://doi.org/10.1111/trf.15619

Prevalence of serologic markers of transfusion and sexually transmitted infections and their correlation with clinical features in a large cohort of Brazilian patients with sickle cell disease

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Background: Patients with sickle cell disease (SCD) often require red blood cell (RBC) transfusion for clinical complications, so may be exposed to transfusion-transmitted infections (TTIs). The prevalence of markers for human immunodeficiency virus (HIV), hepatitis C virus (HCV) and B (HBV), human T-cell lymphotropic virus (HTLV-1/2), Chagas disease, and syphilis in an SCD cohort in Brazil were studied.

Study design and methods: Clinical history, interview data, blood samples, and medical chart review data were collected during cohort enrollment from November 2013 to May 2015. Serologic markers of infection were assessed. Standard measures of statistical association were calculated, and multivariable models were developed for the most prevalent infections to identify associated factors.

Results: Infection markers were evident in 5.2% (144/2779) of the enrolled cohort. Anti-HCV was detected in 69 (2.5%), syphilis antibodies in 34 (1.2%), anti-HTLV-1/2 in 17 (0.6%), HBV surface antigen in 13 (0.5%), Chagas disease antibodies in 13 (0.5%), and anti-HIV in 8 (0.3%) of participants. Factors associated with increased odds of being anti-HCV reactive were older age, illegal drug use, increasing number of RBCs, more than three pain crises in the previous year, and geographic location. Syphilis was associated with older age, females, and smoking history.

Conclusion: HCV infection was more common in older patients who may have received RBCs before testing was performed on donations, suggesting possible historic transfusion transmission. The cohort showed decreasing rates of infections and a reduction in transfusion transmission markers in younger patients compared to historical literature except for syphilis, indicating contemporary reduced risk of TTI.

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Espiritualidade/religiosidade no processo saúde-doença dos pacientes com Doença Falciforme

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Objetivo: Realizar uma revisão sistemática da literatura sobre o impacto que a espiritualidade/religiosidade (E/R) exerce no processo saúde-doença dos indivíduos portadores de Doença Falciforme (DF) e despertar nos profissionais de saúde a possibilidade do uso de medidas alternativas complementares (MACs) associadas ao tratamento convencional, em especial o uso da E/R na DF. Métodos: Foi realizada uma pesquisa com os descritores “spirituality”, “religion” e “sickle cell disease” nas bases de dados eletrônicas SciELO, BIREME, Medline, PubMed e Lilacs no período de 10 a 18 de maio de 2020. Critérios de inclusão: artigos que relacionaram de forma direta ou indireta a E/R aos pacientes com DF. Critérios de exclusão: artigos não pertinentes à relação E/R e DF. Foram selecionados 27 artigos científicos. Resultados: Foi verificado o uso de MACs no manejo dos pacientes com DF com um melhor enfrentamento da doença. A E/R tem alta frequência de utilização e apresentou resultados significativos na qualidade de vida desta população. Considerações finais: O uso da E/R faz parte da atenção integral ao paciente com DF e deve ser acrescido no manejo terapêutico convencional. Os profissionais de saúde encontram dificuldades na abordagem e associação da E/R ao tratamento e sugere-se sua capacitação nesta área.

15- Journal of Advances in Medicine and Medical Research, 32(17): 45-52, 2020.
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Coinfection of Hepatitis B and C with Human Immunodeficiency Virus in Hemophilia: A Cross Sectional Study

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Aims: To identify coinfection by Hepatitis B (HBV), Hepatitis C (HCV) and Human immunodeficiency virus (HIV) in Hemophiliacs. Methodology: We included 107 hemophiliacs. For the current analysis, age, type, severity of Hemophilia and serum profile for Hepatitis B, C and HIV were included. The serological tests performed for Hepatitis and HIV were carried out using the enzyme-linked immunosorbent assay method (ELISA) and the confirmatory tests for HIV and HCV were done by the Western Blot and Polymerase Chain Reaction (PCR). The results of positive and negative serology were compared using the Mann-Whitney test using the Fundação Hemominas reproducibility map of serological reactions, through the CUT-off of each test. Study Design: A cross-sectional study was carried out on hemophilics to asses the prevalence of infections diseases transmitted by transfusion. Place and Duration of Study: Department of Hematology, public service on Coagulopathies at Fundação Hemominas Juiz de Fora Brazil, between January 2008 to December 2018. Results: The average age was 32.77 years with standard deviation (SD)= 16.8 years. In regard to classification, 57.65% of patients with Hemophilia A were severe, and 57% of patients with Hemophilia B were moderate. Laboratory results demonstrated that 24.3% of the patients were HBV positive, and 40.2% were HCV positive, with 21.42% positive by PCR. The prevalence of HIV positive was 11.2%. In general analysis, 44.82% had at least one type of viral infection and 23.4% presented coinfections. HIV positive patients were all positive for HCV (11.2%) and 7.5% of patients were infected by HIV, HCV and HBV. The coexistence of hepatitis C and HIV was statistically relevant with (P = .001), considering the year of birth, most patients with HCV and HIV were born before 1980s (P = .001). Conclusions: Infections by HBV, HCV and HIV are late complications in patients that received blood products before 1990s. Infection by a viral agent and the year of birth has a direct association, due to the standardization and implantation of tests for HIV and HCV in the 80s and 90s.

16- Journal of Integrative Cardiology Open Access, 3(4): 1-3, 2020.
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Situs Inversus Totalis and Sickle Cell Anemia: Case Report and Review of Literature

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Background: Situs inversus totalis (SIT) is a rare, autosomal recessive congenital variation, in which the organs of the chest and abdomen are in a mirrored or reversed position in relation to the usual topography. The incidence of SIT in the general population is low and range of 1 in 8,000 to 1 in 25,000, with the majority of people affected being asymptomatic. Sickle Cell Anemia (SCA), however, is a genetic, monogenic, recessive disease, has a high prevalence in Brazil, with an estimate of 1: 1,350 live births.

Case Report and Methods: The authors report a rare case of SIT in a male child with SCA identified through neonatal screening. The patient's clinical data were extracted from medical records. The diagnosis of SIT was suspected in the pediatric consultation, after altered cardiac auscultation. Screening tests were performed to identify other somatic malformations.

Discussion: SIT is a rare and benign condition, with late and incidental diagnosis in most cases. SCA is a disease diagnosed through neonatal screening with a well-defined multidisciplinary team. In this patient, it was observed that specialized consultations in a reference service for genetic diseases led to early diagnosis of SIT. The importance of this case report is due to the rarity of SIT and the lack of description in the literature of SIT associated with SCA.

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Characterization of HIV risks in a Brazilian sickle cell disease population

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Background: A low prevalence of HIV in sickle cell disease (SCD) patients has been reported in the literature though mechanisms for this are not understood. Methods: HIV risk behaviors were

compared between SCD cases and non-SCD controls using a self-administered audio computer-assisted self-interview. SCD cases were recruited from a multi-center SCD cohort established in Brazil; controls were recruited from SCD social contacts. Categorical variables were analyzed using Chi-Square or Fisher exact test. Continuous variables were compared using the Mann-Whitney U test. Results: There were 152 SCD cases and 154 age/location matched controls enrolled at three participating Brazilian centers during 2016–17. No significant differences in number of sexual partners (lifetime or previous 12 months), male-to-male sex partners or intravenous drug use were observed. Cases received more transfusions, surgeries, and acupuncture treatment. **Conclusions:** Besides the risk of transfusion-transmitted HIV, which is now exceedingly rare, SCD and non-SCD participants demonstrated similar HIV risk behaviors. Causes other than risk behaviors such as factors inherent to SCD pathophysiology may explain the reported low prevalence of HIV in SCD.

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Association of HIV infection with clinical and laboratory characteristics of sickle cell disease

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Background: Sickle cell disease (SCD) is a multisystem disorder characterized by a wide spectrum of clinical manifestations and severity. Studies investigating potential effects of co-morbid human immunodeficiency virus (HIV) and SCD have produced conflicting results, and additional investigations are needed to elucidate whether the interaction between the two disease states might impact both HIV and SCD clinical outcomes. The association of HIV infection with clinical and laboratory characteristics of patients with SCD was assessed.

Methods: This nested case-control study included individuals with SCD with HIV treated at six Brazilian SCD centers. Clinical and laboratory data were abstracted from medical records. HIV

positive participants were compared to age, gender, center, and SCD genotype matched HIV negative participants (ratio 1:4). Individual clinical outcomes as well as a composite outcome of any SCD complication and a composite outcome of any HIV-related complication were compared between the two groups.

Results: Fifteen HIV positive participants were included, 12 (80%) alive and 3 (20%) deceased. Most of the HIV positive patients had HbSS (60%; n = 9), 53% (n = 8) were female, and mean age was 30 ± 13 years. The frequency of individual SCD complications of acute chest syndrome/pneumonia, sepsis/bacteremia, pyelonephritis, ischemic stroke, hemorrhagic stroke, abnormal transcranial Doppler (TCD), and pulmonary hypertension was higher in HIV positive participants when compared to HIV negative, although analyzed individually none were statistically significant. HIV positive participants had significantly higher risk of any SCD complication and of a composite HIV-related complication compared to the HIV negative group (HR = 4.6; 95%CI 1.1-19.6; P = 0.04 and HR = 7.7; 95%CI 1.5-40.2; P = 0.02, respectively). There was a non-significant trend towards higher risk of any infections in participants with HIV positive (HR = 3.5; 95%CI 0.92-13.4; P = 0.07). Laboratory parameters levels were not significantly different in individuals with and without HIV.

Conclusions: In summary, our study in SCD patients shows that those with HIV have an increased risk of any SCD complication and HIV-related complications, as well as a suggestive but not significantly increased risk of infections.

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How ancestry influences the chances of finding unrelated donors: an investigation in admixed Brazilians

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A match of HLA loci between patients and donors is critical for successful hematopoietic stem cell transplantation. However, the extreme polymorphism of HLA loci - an outcome of millions of years of natural selection - reduces the chances that two individuals will carry identical combinations of multilocus HLA genotypes. Further, HLA variability is not homogeneously distributed throughout the world: African populations on average have greater variability than non-Africans, reducing the chances that two unrelated African individuals are HLA identical. Here, we explore how self-identification (often equated with "ethnicity" or "race") and genetic ancestry are related to the chances of finding HLA compatible donors in a large sample from Brazil, a highly admixed country. We query REDOME, Brazil's Bone Marrow Registry, and investigate how different criteria for identifying ancestry influence the chances of finding a match. We find that individuals who self-identify as "Black" and "Mixed" on average have lower chances of finding matches than those who self-identify as "White" (up to 57% reduction). We next show that an individual's African genetic ancestry, estimated using molecular markers and quantified as the proportion of an individual's genome that traces its ancestry to Africa, is strongly associated with reduced chances of finding a match (up to 60% reduction). Finally, we document that the strongest reduction in chances of finding a match is associated with having an MHC region of exclusively African ancestry (up to 75% reduction). We apply our findings to a specific condition, for which there is a clinical indication for transplantation: sickle-cell disease. We show that the increased African ancestry in patients with this disease leads to reduced chances of finding a match, when compared to the remainder of the sample, without the condition. Our results underscore the influence of ancestry on chances of finding compatible HLA matches, and indicate that efforts guided to increasing the African component of registries are necessary.

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Impact of COVID-19 in the attendance of blood donors and production on a Brazilian Blood Centres

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Background: One of the effects of the coronavirus disease 2019 (COVID-19) pandemic is the risk of shortages in Blood Centres. Objectives: To verify the impact of the COVID-19 pandemic on the blood donor's attendance and production of blood components in Fundação Hemominas, a Brazilian public institution was formed by several Blood Centres. Methods: A cross-sectional study was carried out from January to June 2020. Data collected were compared to a historical series from 2016 to 2019. Results: The study showed a reduction in the attendance of blood donors, whole blood collections and blood component production from March 2020, when the first case of COVID-19 was notified in Minas Gerais, Brazil. The results evidenced that Hemominas Blood Centres were affected in a very distinct way by the pandemic with a general mean reduction around 17% in attendance of blood donors and in production of blood components in the period of March to June. On the other hand, the return of blood donors rate increased. Conclusion: The reduction in blood donation during the pandemic period was significant, despite the measures adopted. Still, the recruitment of return donors appears to be an important measure to be considered to decrease the pandemic's effect on blood stocks.

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Association between inflammatory molecules, nitric oxide metabolites and leg ulcers in individuals with sickle cell anemia

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Introduction: Leg ulcers (LUs) are relatively common in patients with sickle cell anemia (SCA). The role of inflammation and nitric oxide (NO) pathways in the pathophysiology of the LU is not understood. Objective: The aim of this study was to verify the association between inflammatory molecules and nitric oxide metabolites (NOx) and the occurrence of the LU in patients with SCA. Method: It was a cross-sectional study on adult participants with SCA followed at Fundação Hemominas, a public blood center in Brazil. Eligible participants were recruited and included in one of two groups: Group 1, comprised of cases with SCA (Hb SS) and at least one LU at the time of inclusion in the study and Group 2, comprised of controls with SCA without a history of LU, matched by sex and age to cases. Participants were interviewed to obtain sociodemographic data and blood samples were collected. Clinical and laboratory data were abstracted from medical records. Nitric oxide metabolites (NOx) and inflammatory molecules were quantified using an immunoassay and Multiplex xMAP® technology, respectively. Eighty-seven individuals were included, ranging in age from 17 to 61 years (mean 40 ± 10.7 years); 30 had LU and 57 were controls without LU. Results:

Participants with LU had significantly higher levels of interleukin 8 (IL-8), IL-10, IL-15, NOx and platelet and white blood cell (WBC) counts, when compared to those without LU. Participants with LU had a significantly higher risk of having a history of osteomyelitis and a higher use of antiseptic soap in bathing, when compared to those without LU. Conclusion: In conclusion, our results showed that NOx, inflammatory molecules and hematological features were associated with LU in Brazilian adults with SCA.

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Cognitive impairment in the HTLV-1 infection: a comparative study associated with functional performance

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Human T cell leukemia virus type-I (HTLV-1) infection courses with a myelopathy, the tropical spastic paraparesis (HAM/TSP). In a case-control study, we compared the neuropsychological profile and functional characteristics in two case HTLV-1-infected groups (asymptomatic and with HAM/TSP) with a control group negative for HTLV-1. Subjects were paired for age, sex, and educational features. The case group differed from control group in neuropsychological measures such as in episodic memory recall, executive functions, and fine motor dexterity measure. Individuals with HAM/TSP have more depressive symptoms and worst performance in activities of daily living (ADL) presenting a less functionality. In multivariate models, the fine motor performance, the executive functioning, the recognition memory, and the depressive symptoms explained part of the variance in functionality. Those findings may contribute to understand of everyday life impairments and limitations of HTLV-1-infected population and to organize the rehabilitation. Once more, based in neuropsychological and functional data, we can reaffirm that HTLV-1 is never a benign condition, but sometimes it is only in a stage coursing with less symptoms.

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Chagas disease: Performance analysis of immunodiagnostic tests anti-Trypanosoma cruzi in blood donors with inconclusive screening results

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Background: The screening ofTrypanosoma cruzi-infected blood donors using two serological techniques frequently leads to conflicting results. This fact prompted us to evaluate the diagnostic performance of four "in-house" immunodiagnostic tests and two commercially available enzyme-linked immunosorbent assays (ELISAs).

Material and methods: One hundred and seventy-nine blood donors, whose screening for Chagas disease was doubtful, underwent three in-house ELISAs, one in-house immunoblotting test (TESA-blot), and two commercial ELISAs (bioMérieux and Wiener) in an attempt to define the presence or absence of infection. Simultaneously, 29 donors with previous positive results from three conventional serological tests and 30 donors with constant negative results were evaluated.

Results: The ELISA-Wiener showed the highest rate in sensitivity (98.92%) and the ELISA-bioMérieux, the highest specificity (99.45%), followed by the TESA-blot, which showed superior performance, with lower false-negative (2.18%) and false-positive (1.12%) rates. In series, the combination composed of the TESA-blot and ELISA-bioMérieux showed slightly superior performance, with trifunctional protein deficiency (TFP)=0.01%.

Conclusion: Our study confirms the high sensitivity and specificity of commercial kits. To confirm the presence or absence of T. cruzi infection, the combination of TESA-blot and ELISA-bioMérieux may be suggested as the best alternative. Individually, the TESA-blot performed the closest to the gold standard; however, it is not commercially available.