Pubmed and Web of Science. The search strategy incorporated keyword combinations related to people with SCD, music, dance, physical activity and health status. The ROBINS-1, Rob 2.0 and Qualsyst tools were used to evaluate the risk of bias.

Results: After the screening process, ten articles were included. Published studies were conducted in Brazil, the USA, France and Saudi Arabia. The total number of participants was 1122 and the age varied between 7-58 years. From all 1122 participants, 550 (49.02%) were women, 407 (36.27%) men and 165 (14.71%) were unspecified. Most studies conclude that exercise and music therapy have positive effects on the health status and quality of life of people with SCD, significantly improving pain intensity and mood. The main finding is that standardized, supervised, and patient-adapted moderate-intensity endurance-exercise training can be safe in patients with SCD without an increased risk for a vaso-occlusive crisis. However, due to heterogenous intervention programs, varying risk of bias and the inclusion of small sample sizes it remains difficult to draw general conclusions. Surprisingly, none of the studies explicitly explored the effect of dance movement therapy in this population.

Summary - Conclusion: To our knowledge, this is the first systematic review to identify the effect of music and dance as forms of physical activity on the health status of people with SCD. Both activities have a wide range of physical and mental benefits and can be a way to stay fit for people of all ages. Further studies with a randomized study design are needed to explore the impact and long-term effects and to compare different forms of musical engagement and dance interventions in individuals. Although multiple benefits have been described, more evidence would motive healthcare professionals to include music and dance therapies as a standard of care in the treatment of people with SCD.

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5588995 COMPARATIVE EFFICACY AND SAFETY OF PHARMACOLOGICAL INTERVENTIONS FOR SICKLE CELL DISEASE IN CHILDREN AND ADOLESCENTS: A NETWORK META-ANALYSIS

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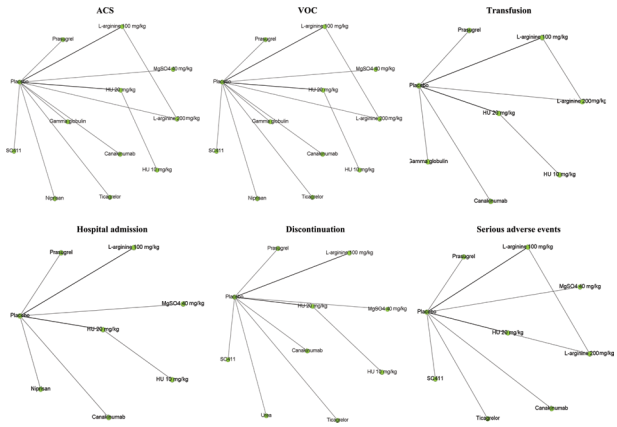
Background: Sickle cell disease (SCD), an inherited hemoglobinopathy characterized by anemia, severe pain, acute chest syndrome (ACS) and vaso-occlusive crisis (VOC), has important impact on morbidity and mortality worldwide, especially in the pediatric population (over 50% die before age of 5). Although few treatment options are available, new disease modifying therapies, intended to prevent or reduce SCD-related complications are under development. Previous systematic reviews are limited to adult patients or focused only on gathering data of few therapies.

Aims: Our aim was to synthetize the evidence on the efficacy and safety profiles of pharmacological interventions for managing SCD in children and adolescents.

Methods: A systematic review with searches in PubMed, Scopus, and Web of Science was performed (May-2022). The protocol is registered at PROSPERO CRD42022328471. We included randomized controlled trials comparing any disease-modifying agent used to treat SCD complications in patients under 18 years old. The outcomes of interest included: VOC, ACS, transfusions, hospital admission, discontinuation, serious adverse events. Data were pooled by means of Bayesian network meta-analyses with surface under the cumulative ranking curve analyses (SUCRA). Results were reported as odds ratio (OR) with 95% credibility intervals (CrI). Additionally, stochastic multicriteria acceptability analyses (SMAA) were performed. The methodological quality of the trials and certainty of evidence were evaluated through RoB 2.0 tool and GRADE approach, respectively.

Results: Overall, 17 randomized controlled trials (n=1,972 patients) published between 1982-2022, conducted mostly in Africa (41%) and North America (35%) were included for analyses. Around one-third of the trials were restricted to homozygous patients for the SCD allele (SS HMZ); yet when reported, patients with heterozygous S-C combination represented less than 30% of the population. Males accounted for 49.0% of the cases, with ages varying from 1-19 years old. Almost all trials (n=15, 88.2%) directly compared active drugs with placebo. The evaluated interventions were: hydroxyurea [n=6 trials], L-arginine [n=3], antiplatelets [n=2], immunotherapy/monoclonal antibodies [n=2], sulphates [n=2], docosahexaenoic acid [n=1], niprisan [n=1] (Figure 1). SUCRA and SMAA revealed that immunotherapy/monoclonal antibodies and hydroxyurea 20 mg/kg are potentially more effective against ACS (17% and 24% probabilities, respectively), VOC (around 29% and 20%, respectively) and needing of transfusions (around 25%), while L-arginine (100-200 mg/kg) and placebo were more associated with these events. Although therapies were overall considered safe, antiplatelet and sulfates may lead to more discontinuations and severe adverse events (uncertainty evidence). Results were similar between age subgroups (<10 years vs. 10-19 years).

Summary - Conclusion: The available evidence on the effect of drugs for managing SCD in children and adolescents is still insufficient and weak. No clear definition and reporting criteria for some outcomes exist. Hydroxyurea 20 mg/kg/day may remain the standard of care for these patients, however, long-term, well-designed trials comparing new immunotherapy/monoclonal antibodies should be performed. The use of monotherapies with L-arginine, antiplatelets or sulphates should be avoided given the poor benefit-risk ratio for this population.



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5613374 ASSOCIATION BETWEEN GENOTYPE AND DISEASE COMPLICATIONS IN EGYPTIAN PATIENTS WITH BETA THALASSEMIA

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Background: In beta thalassemia, the degree of globin chain imbalance is determined by the nature of the mutation of the β-gene. β° refers to the complete absence of production of β-globin on the affected allele. β⁺ refers to alleles with some residual production of β-globin. The homozygous state results in severe anemia that necessitates regular blood transfusion. On the other hand, frequent blood transfusion can lead to iron overload resulting in progressive dysfunction of the heart, Liver as well as multiple endocrinopathies.

Aim: We aimed to evaluate the impact of genotype on the development of disease complications in patients with β thalassemia.

Methods: A Cross sectional study was carried on 73 patients with beta thalassemia. Genotyping was determined by DNA sequencing technique. Routine investigations as well as MRI liver and heart were performed to assess iron overload.

Results: We found that β⁺β⁺ was the most common genotype in our patients followed by β°β° and β°β⁺. Mean Liver iron content (LIC) was significantly higher in β°β° compared to β°β⁺ and β⁺β⁺ genotypes and mean cardiac T2* was significantly lower in β°β° compared to β°β⁺ and β⁺β⁺ genotypes. Hepatic complications, hepatitis C, cardiac