High prevalence of low serum 25-hydroxyvitamin D levels and secondary hyperparathyroidism in Fontan patients: need for increased surveillance of vitamin D status.

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Background. Suboptimal vitamin D status may negatively affect many organ functions. Limited data exist on the vitamin D status in Fontan patients. The aims of this study were to determine the prevalence of low serum 25-hydroxyvitamin D (s25(OH)D) levels among Fontan patients, and to explore potential risk factors for vitamin D deficiency.

Methods. A retrospective chart review was performed of Fontan patients who had been screened for vitamin D status. Vitamin D deficiency was defined as a s25(OH)D level of < 20 ng/ml. The neutrophil-to-lymphocyte ratio (NLR), a marker of systemic inflammation, was calculated in all patients. Associations between laboratory measurements and patient characteristics were explored.

Results. Data were collected from 27 Fontan patients (55.6% male, mean age 8.1 ± 5.3 years). Protein-losing enteropathy (PLE) was diagnosed in 6 patients (22.2%). Mean s25(OH)D level was 14.1 ± 10.4 ng/mL. Vitamin D deficiency was found in 19/27 patients (70.3%), and a severe vitamin D deficiency (< 10 ng/mL) was observed in 12/27 patients (44.4%). s25(OH) D levels were not significantly lower in PLE patients. Patients with increased skin pigmentation had significantly lower s25(OH)D levels (7.9 ± 5.5 ng/mL vs 16.7 ± 5.5 ng/mL, p = 0.04). No further parameters were associated with low s25(OH)D. Hyperparathyroidism was present in 5/21 (23.8%) patients, and was more prevalent in PLE patients (PLE: 4/5 patients, 80.0%; vs non-PLE: 1/16 patients, 6.3%; p < 0.001). Interestingly, circulating parathyroid hormone correlated with parameters of systemic inflammation (NLR: r = 0.484, p = 0.026; relative lymphocyte count: r = -0.635, p = 0.002). Vitamin D supplementation significantly increased s25(OH)D levels (p < 0.0001), and was accompanied by a reduction in parathyroid hormone concentration (p = 0.032).

Conclusions. A high prevalence of vitamin D deficiency was found among Fontan patients, independent of age, time after Fontan procedure, ventricular morphology, and presence of PLE. A potential important link between parathyroid hormone and systemic inflammation is suggested. Further studies are needed to elucidate the clinical role and mechanisms underlying vitamin D deficiency in Fontan patients.