INTRODUCTION: Connective tissue dysplasia (CTD) is a condition in which there is a disruption to the formation of connective tissue in the embryonic and/or postnatal periods. The study reports on the clinical manifestations of cerebral venous insufficiency in children with CTD, and examines the most common features for the timely prevention of complications of the course of the disease. We aimed to better characterize the common features of venous insufficiency symptoms of connective tissue disorders in children.

METHODS: We examined 60 children with signs of CTD and 40 healthy controls in the age group of 10 to 16 years. The inclusion criteria consisted of venous complaints, symptoms, and signs of CTD. Children with hereditary CTD, acquired deformations, trauma or other medical and psychiatric disorders were excluded.

RESULTS: The CTD group, higher levels of total aesthesia (CTD: 45.9 ± 2.89; control: 25.9 ± 3.5) and physical fatigue (CTD: 43.4 ± 3.76; control: 24.9 ± 2.3). The CTD group also scored higher on the CESD depression scale compared to controls (CTD 26.4 ± 2.3; control: 12.3 ± 4.5).

CONCLUSION: Patients with severe CTD require closer attention as early as adolescence. Signs of chronic cerebral venous insufficiency combined with reduced background mood can be predictors of an earlier development of cerebrovascular disease in this group of patients.
METHODS

The connective tissue disorder group (CTD) consisted of 60 children with signs of connective tissue dysplasia aged from 10 to 16 years. The control group (Control) consisted of 40 healthy children.

The connective tissue disorder group (CTD) consisted of 60 children with signs of connective tissue dysplasia aged from 10 to 16 years. The control group (Control) consisted of 40 healthy children. Selection of patients was carried out using the following criteria (above).

Symptoms of connective tissue dysplasia were evaluated by criteria systemic involvement of connective tissue and Scale of hypermobility assessment of joints (Beighton et al., 1988).

RESULTS

The groups studied were comparable in age (average age of 16), gender (male:female ratio CTD 25:49, control 16:10), severity of the condition at the time of admission, and background diseases. The presence of the venous complication symptoms as a percentage of the total group is presented in Table 1.

The CTD group, higher levels of total asthenia (CTD: 45.9 ± 2.89; control: 25.9 ± 3.5) and physical fatigue (CTD: 43.4 ± 3.76; control: 24.9 ± 2.3) were observed. Further, the CTD group scored higher on the CESD compared to controls (CTD 26.4 ± 2.3; control: 12.3 ± 4.5).

DISCUSSION

Children with severe manifestations of connective tissue dysplasia more often revealed signs of cerebral venous insufficiency than children without the disease. Children with severe manifestations of connective tissue dysplasia more often revealed signs of cerebral venous insufficiency than children without the disease.

This can be explained by the extensive damage to organs and systems under the influence of connective tissue dysplasia. Since the circulatory system also includes the presence of connective tissue, the walls of the blood vessels (especially the veins) become extremely vulnerable, resulting in a venous insufficiency.

In children with connective tissue dysplasia more pronounced general and physical fatigue, as well as has a tendency to depressive disorders. Due to the inelasticity and imperfection of the walls of the blood vessels, the blood circulation is disrupted in these patients, and therefore the supply of blood to the main parts of the brain, in particular the limbic system responsible for the centres of mood, attraction and pleasure, is disrupted.

Consequently, hypoxia of such structures could lead to the asthenic syndrome and depression. However, it is necessary to emphasize the complex character of these interactions: Firstly, connective tissue dysplasia is characterized by a combination of multiple features, signs and symptoms. Secondly, there are a huge number of connective tissue anomalies, such as hypermobility of joints, skin striae, epicantbic folds, keeled thorax, among others.

Analysing such complex array of features requires the use of specialised methods to understand the clinical data. At present, a full characterisation of the

INCLUSION CRITERIA

- Venous complaints:
  - headache, dizziness, puffiness of the face and eyelids in the morning; increased occurrence and/or severity of headache, reporting “noise in my head”, visual disorders when wearing tight collars or ties and during sleep with a low headboard, dry eyes (especially in the morning), sleep disturbances (e.g. nightmarish dreams, frequent nocturnal awakenings due to headache), a sense of nasal congestion beyond the symptoms of acute respiratory infections.

- Venous symptoms:
  - swelling of the face / eyelid, redness of the eyes, Venous mesh on the front surface of the chest and/or neck, acrocyanosis, hypoesthesia of 1 or 2 branches of the trigeminal nerve;

- Signs of connective tissue dysplasia assessed according to the published criteria by (Beighton et al., 1988).

EXCLUSION CRITERIA

- Hereditary connective tissue diseases (e.g. Marfan’s syndrome, Ehlers-Danlos syndrome and others)
- Acquired deformations of the musculoskeletal system
- Trauma of the support and movement organs during the last 2 years, accompanied by a violation of the integrity of the bone tissue
- Acute and chronic diseases of internal organs
- Oncological diseases
- Psychiatric disorders

The subjective scale of asthenia 20-MFI (Smets et al., 1995) was used to determine the severity of asthenia. The Center for Epidemiologic Studies Depression Scale (CESD) was used to assess the degree of depressive symptoms (Radloff, 1977).

Statistical analysis was performed using StatSoft Statistica. Paired comparisons were analysed using the nonparametric Mann-Whitney U-test. Differences were considered statistically significant when p <0.05.
risk associated with each feature is not easily accessible or feasible in clinic.

However, an increased understanding of the frequency of the symptoms may lead to an increased awareness and earlier recognition of the complications which may be beneficial for the prevention of further issues to the patient.

CONCLUSIONS

Patients with severe dysplasia of connective tissue require particular attention, as early as adolescence, especially if they have signs of chronic cerebral venous insufficiency, which, combined with reduced background mood, can cause earlier development of cerebrovascular disease in these patients.

In patients with moderate degree of dysplasia, the prognosis is favourable. Children with severe degree of dysplasia should be monitored by a range of specialties, since the changes associated with CTD are of a multisystem nature.

This study has highlighted some of the common characteristics and complaints presented by children with CTD. We plan to study changes in the autonomic nervous system in these adolescents, and conduct an extensive series of studies.

REFERENCES