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AB0919

EFFECTS OF ALEXITHYMIA ON ACTIVITY PAIN, UPPER EXTREMITY FUNCTION, SYMPTOM AND DEPRESSION LEVELS IN PATIENTS WITH THORACIC OUTLET SYNDROME

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Background: Presentation of the most common changes in the thoracic outlet syndrome causing functional disorders of the upper limb (1). The majority of the studies published on TOS highlight physiotherapy strengthening exercises and postural reeducational drills as being the mainstay of any conservative management programme for TOS (2). However, in the literature, the effect of emotional state on progression in treatment programs has not been investigated. Alexithymia, which refers to deficiencies in the self-awareness of emotional states, has been reported to be associated with poor ability in various aspects of social cognition (3). The ability of TOS patients to express themselves and their emotions will affect the success of the treatment.

Objectives: The aim of this study was to investigate the effect of alexithymia on upper extremity functions, symptoms, pain level, depression and anxiety in patient with Thoracic Outlet Syndrome.

Methods: Forty-three TOS patients (36.67±13.99 years; 38 women, 5 men) were enrolled to the study. Alexithymia was assessed with Toronto Alexithymia Scale (TAS-20). The patients were divided into two groups as non-alexithymia group and alexithymia group according to 51 points TAS -20 cut-off score. Pain levels at rest and activity were assessed with visual analogue scale; upper extremity function was assessed with Disabilities of the Arm, Shoulder, and Hand (DASH) questionnaire, and upper extremity symptom was assessed with using Servical Brachial Symptom Questionnaire. Mann-Whitney U test was used for data analyses.

Results: Activity pain level and depression in alexithymia group was higher than the non-alexithymia group, $p=0.008$ and $p=0.007$, respectively. Also, functional level and symptoms were worst in alexithymia patients, $p=0.041$ and $p=0.05$, respectively. No difference was found between groups in anxiety and resting pain level ($p>0.05$).

Conclusion: Thoracic outlet syndrome patients who have alexithymia show worse pain, symptoms, function, and depression. Emotion should be considered in physiotherapy programs. Because the progression of alexithymia patients are worst than non-alexithymia patients.

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AB0920

THE EFFECT OF TIBIALIS POSTERIOR MUSCLE FATIGUE PROTOCOL ON THE STATIC AND DYNAMIC FOOT PARAMETERS

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Background: The tibialis posterior has a important key role as an inverter of the rearfoot. Also providing dynamic support across the midfoot. The value of the tibialis posterior shown by biomechanical research conducted on patients with posterior tibialis tendon dysfunction. One method

of assessing a muscle's contribution to a specific movement pattern is via fatigue-inducing exercise of that muscle. Consequently, to realize the role of tibialis posterior fatigue on foot mechanics it seems prudent to use an exercise that more selectively activates this muscle (1,2).

Objectives: The purpose of this study is to investigate dynamic and static foot parameters changed by tibialis posterior muscle fatigue protocol.

Methods: Thirty patients were participated in this study. Socio-dermo-graphic and descriptive characteristics of the participants were recorded. The changes on the dynamic and static parameters of foot were measured by pedobarography. The two exercises selected for the fatigue protocol are: 1-Unilateral heel raise 2-Close chain resisted foot adduction. Fatigue level was measured by Borg scale. Static and dynamic parameters obtained before and after fatigue protocol were compared.

Results: The mean age of the patients was 24.88 ± 4.39 years, mean height was 171 ± 7.89 cm and mean weight was 68.7 ± 14.5 kg. We found significant increase in the maximum pressure measurement of mid foot ($p=0.004$). In the total contact area during the static position, there were no significant changes ($p>0.05$). Participants' maximal pressure in the middle standing phase was changed from 19.78 ± 10.71 N/cm² to 22.89 ± 13.03 N/cm² after fatigue protocol in medial of middle foot ($p=0.03$).

Conclusion: According to the results, there was a correlation between the fatigue of tibialis posterior and the static and dynamic parameters of foot. The increase in the middle foot peak pressure distributions after the fatigue protocol of the tibialis posterior muscle can be explained as the increase of the angular amount of pronation and prolongation of the time with the middle standing phase. Increase in pressure forces of the middle foot and increase in percentages of the area cause increase of the weight in the front foot during the gait cycle. Thus, fatigue protocol causes similar effects as tibialis posterior muscle dysfunction.

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Paediatric rheumatology

AB0921

JUVENILE-ONSET LUPUS WITH SECONDARY CMV-INDUCED MACROPHAGE ACTIVATION SYNDROME (CASE REPORT)

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Background: Juvenile-onset lupus (JSLE) is a complex illness with an aggressive course and higher chances of morbidities and mortality. JSLE is extremely rare before the age of 5 years. A sole disease criterion may precede long time before others, making diagnosis rather challenging

Objectives: Identifying JSLE and its subsequent complications

Methods: A three and half years old female patient presented with an established diagnosis of idiopathic thrombocytopenia (ITP) of one year duration. One month ago, the patient developed an acute onset of high grade persistent fever, bilateral knee swelling and significant weight loss.

Results: On physical examination, patient was emaciated and pale with multiple ecchymotic patches all over her body. Vital data showed normal blood pressure and apical heart rate ranges for age. Systemic examination revealed multiple oral ulcers, hepatosplenomegaly, bilateral knee arthritis and bilateral below knee painless pitting lower limb edema. Neurological examination was free. Laboratory evaluation showed a CBC with TLC $1.800/\text{mm}^3$ NE $0.5/\text{mm}^3$, HB 7.9 gm/dl, PLT $79 \times 10^3/\mu\text{L}$, ESR 120 mm/1st hr and CRP 96 mg/dl. Routine chemistry showed elevated liver enzymes (ALT 137 IU/L & AST 280 IU/L), LDH 960 U/L, serum albumin 2.5 g/dl, normal renal functions. The history of chronic ITP together with evolution of other autoimmune manifestations raised the suspicion of an underlying Connective tissue disorder. Immunological profile showed positive immune markers (ANA 1/320 speckled and positive Anti-DNA), positive lupus anticoagulant and anticardiolipin IgM, C3 was consumed (49 mg/dl). Secondary Macrophage activation syndrome (MAS)

was suspected and markers revealed serum ferritin 30.000 ng/ml, fibrinogen 250 mg/dl, Triglycerides 350 mg/dl and soluble CD25 12.000U/ml. Bone marrow aspiration confirmed the diagnosis. Cytomegalovirus (CMV) IgM and PCR were significantly high. With regard to all the previous clinical and laboratory features, a CMV-induced secondary MAS in JSLE was confirmed. Patient was managed according to HLH-2004 treatment protocol (pulsed methylprednisolone 30mg/kg/day, cyclosporine A 6mg/kg/day, IV etoposide 150mg/m²) and IVIG. Ganciclovir was also added (5mg/kg IV q12h x7 days). Patient showed marked clinical improvement together with significant diminution of the MAS laboratory markers to normal. Two weeks post discharge on oral dexamethasone and cyclosporine A, the patient showed a severe clinical and laboratory MAS relapse (serum ferritin 50.000 ng/ml). The previous treatment protocol was restarted together with adding Rituximab 375mg/m² x 3 doses two weeks apart. Patient showed marked clinical and laboratory improvement and was maintained on full dose oral prednisone and oral Cyclosporine A. Tight gradual withdrawal of oral steroids was done with close clinical and laboratory follow up.

Conclusion: JSLE is a major diagnostic conundrum in pediatrics owing to its extremely variable clinical manifestations which can mimic many common pediatric conditions (e.g. malignant disease, other auto-inflammatory conditions, malignancy and any specific organ associated disease). Compared to adults JSLE has a more severe disease presentation. MAS is a life threatening condition that requires high index of suspicion and prompt management. In our case, controlling JSLE activity using Rituximab was significantly beneficial in arresting HLH evolution.

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AB0922

UPDATES ON THE EVIDENCE BASED INTERDISCIPLINARY GUIDELINES FOR HENOC-SCHÖNLEIN PURPURA

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Background: Henoch-Schönlein purpura (HSP) is the most common childhood vasculitis, it is characterized by inflammation of small vessels leading to non-thrombocytopenic purpura, arthritis/arthralgia, GI hemorrhage and glomerulonephritis¹. There is a very poor evidence base to guide the management of HSP, particularly for those with the severe forms of HSP nephritis. Up till now there are no EULAR or ACR recommendations for management of HSP.

Objectives: To set an updated interdisciplinary recommendations for the management of HSP

Methods: Interdisciplinary guidelines were developed with representative from PRINTO Egypt for pediatric rheumatology as well as patients' group. A systemic literature analysis in MEDLINE was performed, evidence and recommendations were graded, an algorithm for treatment and final statements were discussed in a consensus meeting (Nominal Group Technique). A preliminary draft was fine tuned and discussed thereafter by all participants (Delphi Procedure).

Results: Consensus was reached on recommendations, including a standardized treatment strategy according to the HSP severity state in the individual patient. In this updated interdisciplinary guideline for HSP; treatment was tailored to the patient's pathology and its severity (figure 1).

Criteria for admission were identified as: severe arthritis or arthralgia limiting ability to weight bearing and mobilization, severe abdominal pain, GI hemorrhage, evidence of nephritis/nephritic syndrome or renal impairment, evidence of neurological symptoms. Criteria for early referral to pediatric nephrologist were also identified including: hypertension, abnormal renal function, macroscopic hematuria for 5-days, nephrotic syndrome, acute nephritic syndrome, as well as persistent proteinuria.

Conclusion: A total of seven evidence based interdisciplinary recommendations for management of HSP have been formulated, that provide an up-to-date guidance of HSP management.

Recommendations for management of HSP

- **Joint symptoms:** bed rest and NSAIDs (avoid NSAIDs if renal disease present). Steroids have also been used in severe cases
- **Abdominal pain** usually settles within 72hrs and simple analgesia is adequate as (Ibuprofen at 5-10mg/Kg/dose tds +/-Paracetamol at 10-15mg/Kg/dose qds)
- **If abdominal pain is severe**, consider using steroids (2mg/Kg/day).
- **If abdominal pain is acute, severe and/or associated with blood in the stool**, then consider Intussusception seek **surgical opinion**. seek surgical opinion also in case of painful scrotal swelling
- **Skin manifestations** are generally benign and self-limited. Severe cases have been treated with Colchicine or dapsone.
- **Treatment of HSP nephritis in children:**
 - Children with HSP nephritis and persistent proteinuria 0.5-1 g/d are treated with ACE-I or ARBs.
 - Children with HSP nephritis and persistent proteinuria > 1 g/d after a trial of ACE-I or ARBs, and GFR > 50 ml/min per 1.73m² be treated with a 6-month course of corticosteroid therapy (either pulse followed by oral steroid 0.5 mg/kg prednisone on alternate days for 6 months or oral prednisone 0.8-1 mg/kg/d for 2 months and then reduced by 0.2 mg/kg/d per month for the next 4 months)
- **Treatment of crescentic HSP nephritis in children** (crescents in more than 50% of glomeruli in the renal biopsy with rapidly progressive renal deterioration):
 - use of pulse steroids and cyclophosphamide and azathioprine
 - Cyclosporine A is effective therapy in children resisted to first line therapy.
 - mycophenolate mofetil could be used in steroid-resistant cases
 - plasmapheresis and IVIG could be used in resisted cases
 - Rituximab could be used in adults with refractory or relapsing HSP but up till now its safety not established in children

Figure 1: recommendations for management of HSP

Figure 1

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AB0923

EFFECTS OF EXON 10 MUTATIONS VS NON-EXON 10 MUTATIONS ON FMF PHENOTYPE AND RESPONSE TO TREATMENT

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Background: Familial Mediterranean fever (FMF) is the most common monogenic periodic fever syndrome. The MEFV gene mutation encoding the pyrin protein results in an uncontrolled increase in interleukin-1. Today, more than 333 MEFV mutations have been identified; however, exon 10 mutations are still seem to be best correlated with clinical findings.

Objectives: In this study, we aim to investigate the role of exon 10 mutations vs non-exon 10 mutations on clinical features and response to treatment in patients with FMF.

Methods: Data charts of children (n=935) with FMF from Dokuz Eylul University childrens' hospital and Dr.B.Uz childrens' hospital were reviewed. Patients were divided into two groups with regard to having