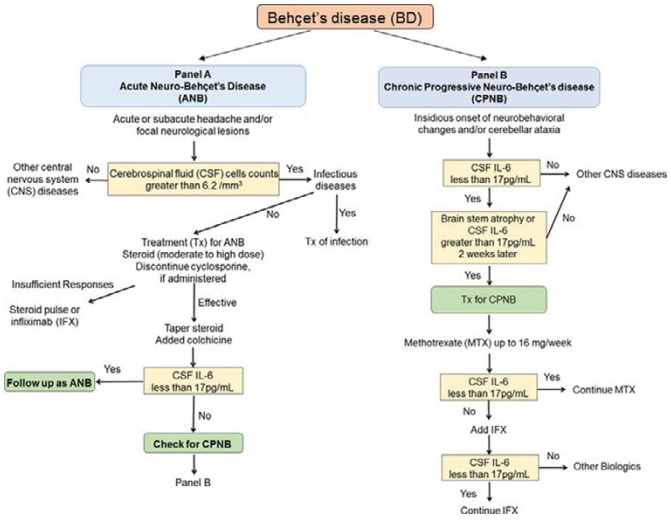
Behçet's disease (NB) and can be classified into acute type (ANB) and chronic progressive type (CPNB) based upon differences in the clinical course and responses to corticosteroid treatment. Diagnostic criteria were generated in 2013 based on a multicenter clinical survey performed by the Behçet's Disease Research Committee of the Ministry of Health, Labor and Welfare of the Japanese Government. Although "Guidelines for Treatment of NB" was also proposed based on the survey, it is still preliminary.

Objectives: The aim of the current study is to develop evidence-based recommendations for the management of NB supplemented by expert opinions where necessary.

Methods: First, clinical questions (CQs) on NB were extracted from a literature search for problem areas and related keywords, and draft CQs and a flow chart were prepared. The expert committee, a task force of the research subcommittee for NB, consisted of 7 board-certified rheumatologists (one was also a board-certified neurologist) and 3 board-certified neurologists. A systematic literature search was performed using Medline and the Japan Medical Abstract Society databases from 1997 to 2016. A total of 15 initial CQs were generated. These yielded the final recommendations developed from 3 blind Delphi rounds, in which the rate of agreement scores on CQs (range 1 [disagree]–5 [strongly agree]) was determined though voting by the whole committee.

Results: Thirteen recommendations were developed for the management of NB (general 1, ANB 7, CPNB 5). The strength of each recommendation was established based on the evidence level as well as rate of agreement. There was excellent concordance between the level of agreement of rheumatologists and that of neurologists. Based on these recommendations, a flow chart was established for the management for ANB and CPNB (Figure).



Conclusions: The recommendations generated in this study are mainly based not only on expert opinions but on the results of uncontrolled evidence from open trials and retrospective cohort studies. Guidelines that can be used for international studies are needed, for which verification by further properly designed controlled clinical trials is required.

Disclosure of Interest: None declared

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FRI0327 GASTROSCOPIC FEATURES AND CLINICAL CHARACTERISTICS IN 172 CASES OF CHILDREN WITH HENOCH-SCHONLEINPURPURA

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Objectives: To investigate gastroscopic features and explore the relationship between clinical characteristics in children with Henoch-Schonleinpuepura (HSP).

Methods: To take gastroscope in 172 cases of children with HSP in our hospital and summarize the gastroscopic performance.All the case were divided into two groups by gastroduodenal mucosal bleeding or not.It was compared among the total time of abdominal pain, pain relief, hospitalization, fasting and kidney injury case in the groups.

Results: Gastroscope with varying degrees of injury of 172 cases has accounted for 169 cases (98.3%).Gastroscopic mainly revealedgastroduodenal mucosal congestion, edema, rough, erosion, bleeding and ulcer, which involved 148 cases of gastric (86.0%), 158 cases of duodenal involvement (91.9%). Mucosal erosion and bleeding occurs mainly in duodenum,mostly in the descending duodenum. Duodenal bleeding accounted for 36 cases (21.8%) in the bulb and 92 cases (55.8%) in the descendant. Only five cases (2.9%) of ulcer occurred in the duodenum, where four cases of bulbar ulcer, one case of descending ulcer. Esophageal and gastric cardia mucosal just occurred in one case. There were not significant difference ($P>0.05$) among the time of abdominal pain, pain relief, hospitalization and fasting in the group. There was no significant difference ($P>0.05$) in the incidence of kidney injury between two groups of children during hospitalization.

Conclusions: Gastroscopic features of HSP in children is characterized by bleeding, erosion of duodenal mucosa and occasional duodenal ulcer formation, which mostly involve the antral mucosa, rarely involving the esophagus, cardia. There was no significant difference ($P>0.05$) among the severity of gastroscopic performance and the time of abdominal pain, fasting,hospitalization and kidney injury of the cases during hospitalization.

Disclosure of Interest: None declared

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FRI0328 THE DIAGNOSTIC VALUE OF ALPHA-1-ANTITRYPSIN PHENOTYPE IN SYSTEMIC VASCULITIS

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Background: Deficiency of alpha-1 protease inhibitor, or alpha-1-antitrypsin (A1AT) is a frequent genetic disorder, which is characterized by low serum level of A1AT and usually manifests as pulmonary emphysema and liver disease. Also the deficiency of A1AT is known to be associated with granulomatosis with polyangiitis (GPA). The influence of A1AT deficiency on GPA clinical course is not clarified.

Objectives: The aim of this study was to estimate the prevalence of pathological A1AT phenotypes in GPA and other systemic vasculitis and to define the influence of A1AT phenotype on clinical course of GPA.

Methods: We enrolled 86 patients with systemic vasculitis, including GPA (N=47), microscopic polyangiitis (MPA, N=16), eosinophilic granulomatosis with polyangiitis (EGPA, N=12), polyarteriitisnodosa (PN, N=11). 46 healthy donors were included in the control group. All blood samples underwent A1AT phenotyping by isoelectrofocusing (IEF) and turbidimetric A1AT measurement. The results of phenotyping were compared to clinical data, such as BVAS activity rate (Birmingham Vasculitis Activity Score), VDI index (vasculitis damage index), organs involvement, inflammatory markers, including antineutrophil cytoplasmic antibodies (ANCA), total IgG concentration and serum levels of C3, C4 complement factors.

Results: Pathological A1AT phenotypes were found in 17% (8/47) of GPA patients, 6,25% (1/16) of MPA patients, 2% (1/46) of healthy donors and were never found in EGPA, PN. The abnormal phenotypes in GPA were 1PiZZ, 4PiMZ, 2PiMF, 1PiMS, and 1PiMS phenotype was identified in MPA patient. Lesion of lung and upper respiratory tract was observed in all patients with pathological phenotypes A1AT (N=8), while in normal phenotype A1AT it was present in 72% and 82% respectively. The mean concentration of A1AT was significantly lower in GPA patients with abnormal A1AT phenotypes, than in patients with normal phenotype A1AT (respectively 1003±148.8 and 1964±127.9 mg/L, $p<0,01$). The average activity by BVAS index in GPA was significantly higher in patients with pathological phenotype A1AT than in patients with normal phenotype A1AT (24,63±2,897 and 18,05±1,444 points, $p<0,05$). Also we revealed excess levels of VDI in cohort of patients with abnormal phenotype A1AT rather, than in cohort of patients with normal phenotype A1AT (6,3±3,1 versus 5,4±2,6, $p<0,05$). The average values concentration of antibodies to proteinase-3 in GPA patients with abnormal phenotype A1AT were significantly higher compared to GPA patients with normal phenotype A1AT (respectively 142,4±25,24 and 86,784±14,98 RU/ml, $p<0,05$). In GPA patients with mutated A1AT phenotypes levels of serum creatinine concentrations ($p<0,01$), levels of total IgG concentration and serum levels of C3 and C4 complement factors ($p<0.05$) also were significantly higher than in group of GPA patients with normal A1AT phenotype.

Conclusions: Pathological A1AT phenotypes are more often observed in GPA patients who have more severe GPA clinical course and higher immunological disease activity.

Disclosure of Interest: None declared

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FRI0329 EFFICACY AND SAFETY OF INFLIXIMAB ORIGINATOR IN PATIENTS WITH TAKAYASU ARTERITIS WITHIN THE RTU (TEMPORARY RECOMMENDATION OF USE) IN FRANCE

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Background: The benefit/risk ratio of infliximab in refractory patient with Takayasu arteritis (TA) is assumed to be favorable, based on retrospective studies with limited sample size [1, 2] in which infliximab has been prescribed off-label.

Since 2013, the French Temporary Recommendation of Use (RTU) provides a temporary framework allowing the use of infliximab originator in "TA patients refractory to conventional treatment" during a 3-year period.

Objectives: The aim of the study was to evaluate the real-life efficacy and safety of infliximab originator in TA patients initiating or with ongoing infliximab treatment.