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P1615 ASPECTS OF THE THERAPY SELECTION FOR IMMUNE THROMBOCYTOPENIA IN CHILDREN

Topic: 32. Platelet disorders

Sviatlana Khoduleva*¹, Irina Novikova¹, Alla Demidenko², Ирина Ромашевская², Ekaterina Mitsura², Oksana Zhuk², Elena Borisova²

¹Internal Diseases Department, Gomel State Medical University, Gomel, Belarus; ²Republican Research Center For Radiation Medicine And Human Ecology, Gomel, Belarus

Background:

Immune thrombocytopenia incidence ranges from 1.6 to 5.6 per 100,000 per year. The level of platelets requiring immediate initiation of therapy is a debatable issue in each case. The choice of the optimal regimen for both the first and second lines of ITP therapy remains an urgent problem. The treatment of chronic continuously relapsing forms of the disease is of particular difficulty.

Aims:

Assessment of the effectiveness of various regimens for the treatment of immune thrombocytopenia in children.

Methods:

262 patients with primary ITP aged from 2 months to 18 years were included into the study. The selection of initial therapy was carried out taking into account the criteria for predicting the chronic course, developed based on the results of the previously obtained clinical and laboratory characteristics of various variants of the course of ITP. The following were considered as predictors of a chronic course: the duration of the pre-diagnostic period of more than 17 days, the absence of a possible etiological factor, hemorrhagic syndrome of mild severity, age over 10 years, decrease in cytotoxic CD8-positive cells. As initial first-line therapy, 182 patients (69.4%) received glucocorticosteroids at a standard dose of 1–2 mg/kg of prednisolone. In the absence of criteria for predicting a chronic course, the therapy selection included the administration of intravenous immunoglobulin (IVIG) 1 g/kg once or at a dose of 0.5 mg/kg as monotherapy or in combination with solumedrol pulse therapy at a dose of 15-30 mg/kg per day. Efficacy was evaluated according to WHO criteria (2009).

Results:

A complete clinical and hematological response to first-line therapy by glucocorticoids at a standard dose was obtained in 89%, chronicity was noted in 24%. When predicting an acute course and conducting monotherapy with IVIG, pulse therapy with methylprednisolone, a clinical and hematological response was obtained in 80.4% of patients, however, chronicity was observed much less frequently and amounted to 15%. Therapy for chronic ITP recurrence was carried out with a decrease in platelet count less than 50×10^9 /l and the presence of hemorrhagic syndrome. IVIG was administrated, four-day courses of dexamethasone at a dose of 0.5 mg/kg/day intravenously up to 4 courses, interferon-alpha, pulse therapy with melitprednisolone intravenously. Responses to therapy were observed in 84% of cases, but the duration of remission achieved ranged from 2 weeks to 6 months. Interferon-a therapy at low doses was performed in 9 patients as second-line therapy. A complete response was obtained in 7 patients (77.8%).

Revolade (Eltrombopag) at a daily dose of 25 mg was administered to two patients with continuously relapsing chronic ITP resistant to first-line therapy. Clinical and hematological response was received. Splenectomy in the observation cohort was performed in 12 patients (4.6%) with a chronic continuously relapsing course (14%) and resistant to first and second line conservative therapy. The effectiveness of splenectomy is 86%.

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Summary/Conclusion:

The optimal method of primary therapy for ITP in children with predictors of a chronic course is glucocorticosteroid therapy at a standard dose of prednisolone 2 mg/kg with an optimal duration of no more than 2 weeks. In the absence of predictors of a chronic course in patients, as well as in children under the age of 3 years, it is advisable to use IVIG as monotherapy or combined with methylprednisolone pulse therapy. It is necessary to use thrombopoietin receptor agonists more widely, as well as interferon medicine in modified doses in steroid-resistant patients, which will allow avoiding splenectomy, which is extremely undesirable in childhood.

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