DIFFERENTIAL DIAGNOSIS OF HAEMORRHAGIC DIATHESIS IN CHILDREN

(BY THE MATERIAL OF II-ND CHILDREN'S CLINIC
OF THE HIGHER INSTITUTE OF MEDICINE — VARNA)

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The disturbances of human homeostasis are rather frequent in children. By the material of the II[nd] Children's Clinic in Varna the diseases of homeostasis take second place after anaemias when blood diseases are considered.

A total of 275 children with haemorrhagic diathesis were hospitalized, examined and treated in the clinic. There were 159 boys (63,3 per cent) and 116 girls (36,7 per cent). According to the kind of the haemorrhagic diathesis their distribution was as followed: coagulopathies — 37 children (14,8 per cent), thrombocytopenias — 73 ones (29,2 per cent), and capillarotoxicosis (M. Schönlein'Henoch) — 165 ones (56 per cent). According to the sex the distribution of the patients was as followed: concerning coagulopathies there were only boys while concerning thrombocytopathies there were 41 boys (53,2 per cent) and 32 girls (46,8 per cent). Males: females ratio was nearly one concerning capillarotoxicosis: 81:84, resp. 49,6 per cent: 50,4 per cent.

Haemophilia type A (VIII[th] factor deficit) was found out with 36 from a total of 37 children with coagulopathy. One child had Willebrand-Jurgens' disease. Earliest hemophilia signs were observed in a 43-days old boy with the main manifestation of a recidivans intracranial haemorrhage. The disease was diagnosed during the first year of life in 7 children (19 per cent), between 2 and 3 years after birth — in 10 ones (27 per cent); in the age from 4 till 7 years — in 15 ones (40,5 per cent) and in that from 8 till 14 years — in 5 children only (13,5 per cent).

Injury bleeding is the initial clinical symptom. In children up to 3 years old it is very difficult or even impossible to establish the provoking moment. Most commonly, haemorrhages in the skin, hypoderma and muscles as haemophilia manifestations can be seen (one or even several haematomas). In 2/3 of the cases upper and lower extremities are affected. The hemorrhages are located in the lumbo-sacral and inguinal regions but rather rarely on the trunk. The articular localization of haemorrhages comes next in frequency. However, haemarthroses are most long-lasting and cause severe disability of the children. During the first hospitalization haemarthroses can be found in one third of the children most frequently of the knee joints. The following recidives of the illness are characterized with the presence almost always of recidives of one and the same joint — knee, talocrural, elbow and iliac joint. At the beginning haemarthrosis is monoarticular but then symmetric haemarthroses, chiefly gonarthroses, can be seen. Tongue bleeding is observed in 3 infants (8,1 per cent), abdominal. retroperitoneal haemorrhages — in one child (2,7 per cent), brain haemorrhage — in another one child (2,7 per cent) but haematuria — in 5 ones (13,5 per cent). Haemophilia has a cyclic course due to recidives.

Concerning thrombocytopenias only 3 children were up to 1 year old (4,1 per cent). 14 children were up to 3 years old (19,2 per cent); 29 ones — up to 7 years old (39,7 per cent), and 27 ones — up to 14 years old (37,1 per cent). Two children were with M. Glanzmann's thrombasthenia.

The disease began acutely one-two weeks up to one month before hospitalization in 54 per cent of the children but more than 1 month — in 46 per cent of them. In the clinic all the children have a haemorrhagic syndrome — petechiae were seen in 96 per cent, both petechiae and echimoses in 74 per cent, epistaxis combined with petechiae in 38 per cent, oral mucosa bleeding in 42 per cent, and brain haemorrhages in 3,9 per cent (3 patients) of the children. More rarely suffusiae, suggilatiae, single haematomas and haematuria were also observed.

The anamnestic data showed that more frequently (in 2/3 of the ccases) the initial symptoms were suffusiae on thighs and calves. They were single and without any clear reason at the onset of the illness. They became more frequent, the number of the haemorrhages increased and they appeared at places which were injury-protected what suggested to us that they could be due to thrombocytopaenia. An anaemia is commonly not present. The spleen is enlarged in 50 per cent of the cases. The etiologic factor can't be always established. Concerning our patients the direct causative factor of the disease could not be found out in 54 per cent of the patients. In other cases the illness is preceeded by an acute catarrh of the upper respiratory tract, by infectious diseases (rubeola, varicella, drug administration — ampicilline, analgin, bayrena).

Concerning Schönlein-Henoch's disease only 2 children were younger than 3 years (1,5 per cent). There were 77 children in the age group between 4 and 7 years (46,2 per cent) and 86 ones (52,3 per cent) in the age group up to 14 years. There are some seasonal changes concerning hospitalization of the patients with Schönlein-Henoch's disease only. In spring 43,3 per cent of the children and in autumn 26,1 per cent of them were hospitalized. Commonly, typical symptoms appeared after a catarrh of the upper respiratory tract (in 68,1 per cent of the cases), next came scarlet fever, anti-cholera vaccine application, consumption of certain foodstuffs, sulfonamide administration (a total of 10,9 per cent). The causative factor was not found out in 21 per cent of the cases.

A characteristic peculiarity of the illness is its acute onset in all children studied- high temperature, weakness, and an indisposition. There was an articular syndrome with rapidly transitory pains or with joint oedema, symmetric affecting of the joints, most frequently of the talocrural, elbow, radiocarpal and knee ones, in 75 per cent of the cases. However, articular lesions did not persist. The diagnosis could be made according to the typical exanthema — papulo-haemorrhagic rash — in 100 per cent of the patients. The rash can be scanty, abundant, symmetric and affect most intensively the extensor surfaces. It always appears on calves, more rarely on forearms, on elbow joints, on gluteal regions, auriculae and nose. More rarely, necrotic plaques and blisters filled with haemorrhagic fluid as well as angioneurotic oedema could be observed. There as a certain dependence of rash intensity upon orthostatic position. The appearance of haemorrhages has a cyclic character.

An abdominal syndrome (abdominal pains and melaena) was established in 78 per cent of the cases. A macroscopic haematuria combined with a severe skin and abdominal syndrome was observed in 23 per cent of the children. There were renal disorders in 56,6 per cent of the infants. Only with the children with severe abdominal syndrome a slightly or even stronglier expressed anaemia was established.

The laboratory investigation showed that bleeding time became longer up to 15—20 min in haemophilia children. No retraction even occurred because blood retained as fluid. The thrombelastogram indicated as followed: r+k time — between 20 and 90 min; recalcification time — 300—400 sec, and heparin tolerance over 400 sec. There was also an anaemia (Hb rates between 11 and 6 g %) in 55 per cent of the children. Bleeding time became longer in 58 per cent of the children with thrombocytopaenia with a mean value of 7,7±6,5 min. Thrombocytes were reduced in all children. They reached 50 000 in 88 per cent and even 10 000 — in 40 per cent of the cases as determined pro cmm. Blood thromb retraction was longer than the normal rate in 88 per cent of the cases. TEG maximal amplitude decreased from the normal rate by 10—42 mm. Microscopically the following changes were observed: predominant microthrombocytosis (45 per cent), macrothrombocytes — heavy thrombocytes, located separately and stained basophylically — 25 per cent. There was a typical ultrastructural picture of thrombocytes in children with Glanzmann's disease — lack of aggregation, macrothrombocytes.

In 99 per cent of the children with Schönlein-Henoch's disease bleeding time, coagulation time, TEG and thrombocytes were within the normal ranges. There was a reduction of thrombocytes count from 60 000 till 15 000/ccm, and of fibrinogen from 105 till 45 mg % together with positive protamine and ethanol test.

Discussion

According to our data the allergic vasopathies are the most frequent haemorrhagic diatheses in children followed by thrombocytopaenias where the same mechanism is obviously related to unlocking of the illness (4, 5). Excepting haemophilia that is a lamentable privilege of the male sex the rest haemorrhagic diatheses in childhood did not demonstrate any sexual predominance (1, 6, 8). In the polyclinic the following mistakes of examination were made: when only epistaxis is present the varices of nasal septum are considered only, general diseases such as hypertension are overlooked. The characteristique of the rash is often neglected and local drugs are administered to treat the anaphylactoid purpura.

The laboratory investigations show that coagulation time is a reliable index only in cases with congenital coagulopathies. Bleeding time changes after a significant thrombocyte count reduction below 50 000/cmm.

Both microscopic and electron microscopic studies show that not only in Glanzmann's disease but also in any other thrombocytopaenias functional disorders of thrombocytes can be established. This fact is confirmed by studying the aggregation and adhesion of thrombocytes with prostaglandin, collagen, etc. (2, 3).

The old concept that no changes of plasma and thrombocyte coagulation factors set in in patients with Schönlein-Henoch's disease (1, 4, 5) is already shaked, indeed. The presence of the syndrome of disseminated intravasal coagulation is sure and it is supported by the investigations of the heparin effect when applied in these cases.

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ДИФФЕРЕНЦИАЛЬНЫЙ ДИАГНОЗ ГЕМОРРАГИЧЕСКИХ ДИАТЕЗ У ДЕТЕЙ (НА МАТЕРИАЛЕ II ДЕТСКОЙ КЛИНИКИ ВМИ — ВАРНА)

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РЕЗЮМЕ

Проведено клиническое и лабораторное исследование 275 детей (159 мальчиков и 116 девочек), лечившихся по поводу геморрагического диатеза во II детской клинике. В зависимости от вида заболевания больных разделили на три группы: с коагулопатиями — 37 детей (14,8 %), с тромбоцитопениями — 73 ребенка (52,2 %) и с капилляротоксикозом (М. Шонлайн-Генок) — 165 детей (56 %). Цифры показывают, что по частоте первое место занимают аллергические вазопатии, при которых аллергические моменты связаны с началом заболевания. Наши исследования позволили сделать вэвод, что диагноз «геморрогический диатез» можно поставить в случаях охвата более обширных областей кожи, а также в случае их рецидивирования, их прогрессивного развития, когда они охватывают по крайней мере две области и сочетаются с соответствующей лабораторной констелляцией.

Использование указанных клинико-лабораторных критериев обеспечивает провильную постановку диагноза даже и в амбулаторных условиях.