

HAEMOPHILIA IN BACĂU COUNTY

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Abstract: It has been made a research by comparison of the cases of haemophilia at infants from Bacău county. During 1998 – 2003 6 cases of haemophilia were discovered amongst a lot of 27,623 subjects that have been investigated. The 6 cases of haemophilia found and confirmed are distributed as it follows: 3 cases of haemophilia type A, 2 cases haemophilia type B and one case of vascular haemophilia. These cases are children of age over 10 years old and who come mostly from the urban environment.

INTRODUCTION

Haemophilia is a trouble of coagulation due to some deficiencies of the VIII factor and/or IX and XI factors of the coagulation. The factors of coagulation – plasmatic proteins – interfere and obstruct the wound in the case of an affected blood vessel. Any abnormality and the quality or quantity of these factors leads to haemophilia. The following types of haemophilia are known:

- Haemophilia type A is a trouble in coagulation due to the chromosome X and a deficiency of the VIII plasmatic factor. Its incidence is 1/5,000 – 10,000 of male newborns and on female newborns are very rare cases;
- Haemophilia type B is a trouble in coagulation due to the chromosome X and a deficiency of the IX plasmatic factor. It is from 5 to 10 times less frequent then the other form (WALTER – ROȘIANU, 1986);
- Haemophilia type C is a trouble in coagulation due to the deficiency of the XI plasmatic factor and it can be transmitted autosomal recessive;
- Vascular haemophilia (VON WILLEBRAND disease) is due to the abnormalities of the VIII factor, adding a slight deficiency of the IX/XI factors. Its transmission is autosomally dominant (GEORMĂNEANU, 1986).

MATERIAL AND METHODES

During 1965 – 1975 in Bacău county were discovered 18 cases of haemophilia from which 16 haemophilia type A and 2 cases haemophilia type B from a lot of 60,000 subjects. In order to discover these cases it has been applied the clinical examination in order to explore: the plasmatic and thrombocytic factors, blood investigations – haemoleukogram and the determination of the blood group, testing the sight for distinguishing the colours, radiological investigations regarding the bone system, but also doing the family investigation in order to realise the genealogical tree (ANTON, 1975).

After 30 years, during 1998 – 2003 a number of 6 cases of haemophilia were found, by investigation a lot of 27,623 subjects represented by children with ages between a couple of days and 16 years old. These patients have been discovered after they have suffered some injuries and the paraclinical examination has confirmed the existence of haemophilia. The diagnosis of certainty was confirmed after the determination of the VIII or IX factor of coagulation (ȘTEFĂNESCU, 2004). After studying the background (pedigree) of the family, the existence of some maternal relatives who had different types of haemophilia was detected.

RESULTS AND COMMENTS

The 6 cases of haemophilia discovered during 1998 – 2003 (table 1) are distributed as following: 3 haemophilia type A, 2 haemophilia type B and one case of vascular haemophilia.

Table 1 - The frequency of haemophilia in Bacau county

Year	Number of subjects	Cases of haemophilia	The frequency haemophilia (%)
1998	5183	-	-
1999	4878	1	0,205
2000	4819	2	0,415
2001	3919	1	0,255
2002	4413	-	-
2003	4411	2	0,453
1998 – 2003	27623	6	0,217

The ratio between haemophilia type A and haemophilia type B is 3/2 and in medical literature the ratio is 10/1 (BOWEN, 2002). In the researches made during 1965 – 1975 were discovered 18 cases of haemophilia from a lot of 60,000 subjects in a period of 10 years. The increased number of cases is due to the legislation from that period which has forbidden the interruption of pregnancy and the impossibility of making general genetic tests for confirming the disease. So, the ratio between haemophilia type A and haemophilia B was 8/1 (16 cases type A/2 cases type B) and one case of vascular haemophilia.

From 6 cases of haemophilia, 5 cases are boys and only one case is a girl of 11 years old from Bacău who has vascular haemophilia (table 2). The diagnosis of this girl was put after a metroragy, then she was sent to “Fundeni” Hospital București. The disease has all three characteristics: the deficiency of the VIII factor, low number of thrombocytes and the defect of the capillary vessels. The girl had her skin and the mucous membranes bleeding when she was slightly wounded, on epistaxis and metroragy.

Table 2 - The distribution and the frequency of the cases of haemophilia on sexes

Year	Cases of haemophilia	The number and the frequency of cases (%)			
		girls		boys	
1998	-	-	-	-	-
1999	1	-	-	1	100
2000	2	-	-	2	100
2001	1	-	-	1	100
2002	-	-	-	-	-
2003	2	1	50	1	50
1998 – 2003	6	1	16,66	5	83,34

The cases of haemophilia are composed of children of the age over 10 years old (table 3). These children were discovered after they have suffered some injuries or removed teeth when it was noticed that the time of bleeding was increased (RUSEN, 2004) more than it would be in normal conditions.

Table 3 - The distribution and the frequency of haemophilia according to the groups of ages

Year	Number of cases	Groups of ages and their frequency (%)									
		0-1 year	1-3 years	3-5 years	5-7years	7-10 years	over 10 years				
1998	-	-	-	-	-	-	-	-	-	-	-
1999	1	-	-	-	-	-	-	-	-	1	100
2000	2	-	-	-	-	-	-	-	-	2	100
2001	1	-	-	-	-	-	-	-	-	1	100
2002	-	-	-	-	-	-	-	-	-	-	-
2003	2	-	-	-	-	-	-	-	-	2	100
1998 – 2003	6	-	-	-	-	-	-	-	-	6	100

In the researches made during 1965 – 1975, the children with haemophilia were of different ages as following:

- the group 0-1 year – one case;
- the group 1-3 years – 3 cases;
- the group 3-5 years – 4 cases;
- the group 5-7 years – 3 cases;
- the group 7-10 years – 5 cases;
- the group over 10 years – 2 cases (ANTON, 1975).

At the same time, the children with haemophilia were from urban environment: Comănești – 6 cases and Bacău – 2 cases. In the rural environment, the distribution of the cases with haemophilia type A was the following: Urechești – 3 cases and Huruiești, Plopana, Colonești, Zemeș and Solonț – one case. The cases with haemophilia type B were in: Onești town – one case (urban environment) and Sănduleni – one case (rural environment).

In the researches made during 1998 – 2003, the children with haemophilia are coming especially from the urban environment – 4 cases from which 3 are from Bacău city and one case from Moinești town. In the rural environment were 2 cases: one case in Filipești and the other from Agăș. The cases with haemophilia type A are coming from Moinești – one case and Bacău – 2 cases. The cases with haemophilia type B are coming from Filipești and Agăș both with one case and vascular haemophilia from Bacău city (table 4).

Table 4 - The frequency of the cases of haemophilia in rural and urban environment

Year	The number and the frequency of cases (%)					
	Urban environment		Rural environment		Total cases	
	Bacău	Other cities				
1998	-	-	-	-	-	-
1999	1	100	-	-	-	1
2000	-	-	1	50	1	50
2001	1	100	-	-	-	1
2002	-	-	-	-	-	-
2003	1	50	-	-	1	50
1998 – 2003	3	50	1	16,67	2	33,33

In 4 of 6 cases of confirmed haemophilia some complications appeared due to the bleeding with different locations (table 5). Three cases with complications are represented by the boys with haemophilia type A and type B who have haemarthrosis and blood accumulation on the right knee, the left elbow, the right arm and anaemia post haemorrhage. Another case with complication is of the girl with vascular haemophilia. When this case was discovered, the girl had metroragy and anaemia due to this haemorrhage.

Table 5 - The frequency of cases of haemophilia with complications

Year	Cases of haemophilia	The number and the frequency of cases with complications (%)	
1998	-	-	-
1999	1	-	-
2000	2	1	50
2001	1	1	100
2002	-	-	-
2003	2	2	100
1998 - 2003	6	4	66,66

In medical literature, the severe forms contain cephalic haematoma or extended haemorrhage at the level of the navel cordon (SANDOVICI, 2004). The children with haemophilia during 1965 – 1975 had many bruises, haemarthrosis, epistaxis, bleeding gums and faeces mixed with blood.

At present, there weren't registered any deceases among the children with haemophilia. Thirty years ago there were 2 deceases among the patients of less then one year old who had meningocerebral haemorrhage and urine with blood (ANTON, 1975).

Nowadays, the haemophilia doesn't cause victims anymore, because of the treatment which consist of a transfusion of blood which has embodied the missing factor of the coagulation of blood, also of the increased possibilities of liver transplant and genetic therapy (RAICU G., 2004), but in the same time of the increased possibilities of genetic examination and advice.

CONCLUSIONS

The researches in the cases of haemophilia at infants from Bacău county in two different periods of time revealed the following:

During 1965 – 1975 were discovered 18 cases of haemophilia more than 6 cases in 1998 – 2003.

Compared to the previous period, in present the frequency of the cases of haemophilia type B was increased, also it has been discovered a case of vascular haemophilia. All these cases have a positive family pedigree.

The children with haemophilia are over 10 years old and the most of them are boys; only one case is represented by a girl of 11 years old who has vascular haemophilia, case that has been confirmed by "Fundeni" Hospital București.

The discovered cases are coming from the urban environment, from which Bacău city has 50% from the cases of infants with haemophilia.

Even if the actual progress of science allows that the result and the recovery to be made immediately after the local treatment and transfusions of blood which has embodied the missing factor of the coagulation of blood, it also must pay attention to the prevention of this disease based on the examination and specific advice.

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