

Adaptation in Hemophiliac Adolescents

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■ Mattsson and Gross have discussed the "coping mechanisms" which are used in successful adjustment to hemophilia in boys and young men². In their study of 35 hemophiliac boys and young adults, 27 were considered to have adapted well at school, in the home, and with their peers, with few limitations except for those realistically imposed by the disease. In those instances of failure, difficulties seem to fall into the category of behavioral disturbance with either excessive risk-taking behavior or inappropriate passivity.

However, an investigation of the social and psychological adjustment of 26 hemophiliacs at this center³ revealed a high incidence (46%) of gross sexual and marital maladjustment, in spite of a low frequency (23%) of frank psychiatric problems. The ten adolescent patients in this group were studied more closely to identify precursors of the sexual and marital maladaptations, and a variety of problems were found which make us consider the adaptation of virtually all these adolescents to be sub-optimal. Although the overall educational achievement was impressively high in the younger group, several instances were found of inhibition of school performance going beyond the interference of bleeding episodes. In some cases adjustment at school, in the family, and with peers was superficially good, but closer examination revealed areas of considerable conflict and impairment. These difficulties can be loosely categorized into the following areas:

1. Problems in vocational and scholastic achievement, involving the child's refusal or reluctance to attend school, learning difficulties, or a seeming lack of goals and ambitions.

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2. Behavior problems, including vandalism and other forms of delinquency.

3. A tendency towards accidents and injuries, often the result of reckless activities.

4. Social or sexual maladjustment, as evidenced by inability to relate to members of the opposite sex or a very ambivalent attitude towards them.

The thesis of this paper is that in young hemophiliacs the behavioral disturbances, learning difficulties and sexual disturbances can be related to the failure to resolve universal developmental issues which are made particularly critical by the nature of hemophilia and its manifestations. The main issue is the achievement of stable masculine identification. Because of (1) enforced passivity, (2) difficulties in achieving autonomy at the appropriate age and (3) heightening of castration anxiety, there is a tendency towards excessive identification with the mother who is also the source of the hemophilic genes. The persistence of dependency, lack of age appropriate autonomy, and inability to overcome the excessive castration anxiety all result in feelings of guilt, helplessness, and inability to adequately channel aggression into constructive uses in play and in work. Attitudes toward the opposite sex may be too guilt-laden for adequate sexual interest or they may be characterized by exploitative and overly aggressive feelings.

REPRESENTATIVE CASE REPORTS:

Difficulties in School Achievement

Case 1. L.K. is a 17-year-old boy who was discovered to have a bleeding disorder at 18 months when he had swelling of his knees. He has had frequent hospitalizations, mostly for hemarthrosis in the knees, and has some residual impairment. He uses crutches or a cane much of the time.

In the hospital, he has been anxious, demanding, tearful and at times uncooperative. He was especially fearful of doctors putting needles into his joints. He appeared younger than his age and tended to speak only when spoken to. His speech was excessively intellectual and he used polysyllabic words when he could. Sometimes he became tearful without apparent cause. When he was with his mother he would try to turn questions to her and take a back seat.

Mrs. K. is a competitive, aggressive female who has already outdone her husband in educational achievements and continues in graduate school in order to obtain a doctorate. This has involved long separations from her husband and L., her only child. Her aspirations for L. are equally high, to be a lawyer, teacher, or engineer.

Mrs. K. welcomed the opportunity for L. to be seen by a psychiatrist and psychologist because she wanted to "have him re-tested" to see whether his I.Q. was really lower than it was supposed to be. L. had been having trouble with schoolwork, and took little interest in school except at exam times. He was contrite about his "mediocre" grades, even though there were some A's and no failures. L. reflected his mother's aspirations but doubted his abilities to get into college.

L.'s father stayed in the background. He seemed warmer than Mrs. K. but fearful of L.'s physical activity. His background was that of an Army sergeant and skilled construction worker. L. complained that his father left him only "women's work" to do, cooking and cleaning, while Mrs. K. was away. In discussing this, L. tearfully described putting his fist through a plaster wall. His feeling was one of helpless rage.

In the interview L. revealed that he was preoccupied with fantasies which interfered with his schoolwork. For example, he told a story which was partly borrowed from a novel: A slave marries the wife of a cruel plantation owner after the latter is killed in the Civil War. The master was really not a bad man.

The former slave is unable to make a go of the plantation and it "goes to seed".

L.'s frustration in his attempts to mature can be understood on the following grounds: 1) Mother's ambivalently over-ambitious and possessive attitudes toward him. There is a history of bleeding on mother's side of the family. It is of great importance that two years before L.'s birth, a boy was born who died a few days later of unknown causes. L., the only child, has been "the center of my life", according to his mother. One can be reasonably sure that the loss of the first male child and subsequent discovery of L.'s condition have been deeply injurious to Mrs. K.'s narcissism and her capacity for unambivalent mothering. It may have determined the decision to have no more children. Mrs. K. depreciates L.'s schoolwork as unchallenging and "easy". In spite of suggestions that his learning difficulties are not intellectual in nature (his performance on intelligence tests is in the "very superior" range), and that L. is already too critical of himself, Mrs. K. continues to insist on further achievement and intelligence testing. 2) Father's fears and rejection of roles for L. which involve a measure of risks, even though they might add greatly to L.'s self-esteem, e.g. driving a school bus. 3) The actual interference of recurrent hospitalization and crippling on his schooling, physical activity, peer relationships and on his own concept of being "normal" and acceptable.

In the patient's story it is not hard to see his identification with the guilty slave who gets revenge on father for making him do "women's work", only to fail in his attempts to compete. The bleeding disorder is for him the "curse" or punishment that has been laid in him and his progeny. The fact that there is a real defect in his "seed" makes the resolution of the persistent oedipal conflict much more difficult.

Case 2. Q.V. is a 15-year-old boy with severe vascular hemophilia manifested at circumcision shortly after birth. About the time he was learning to walk, he fell with a fork

in his mouth, suffering a throat injury which required transfusions and hospitalization. Currently he is in eighth grade having lost one year of school due to illness in the third grade. His teachers assert he has a high I.Q. but refuses to work. He seems to show interest only in sports, including touch football.

The patient's older sister, who is in good health, is doing very well in school in contrast to the patient. Mother, who is the more educated parent, is highly oriented towards intellectual achievement. Father has only an eighth grade education and is a maintenance man who enjoys his work but feels his son would not be able to do the rough tasks that he does. The parents agree that Q. shows sullen and rebellious behavior at school and also towards the maternal grandmother who is overly solicitous towards him. He rejects any concern or attention from doctors and nurses. He denies the presence of any vulnerability and resents being prevented from playing tackle football.

The patient appeared hostile and resentful during the interview. He denied the existence of any bleeding disorder and answered questions in monosyllables. Mother dominated the discussion and father took a back seat. Q.'s lack of interest and failure to achieve in school probably reflect an attempted identification with his poorly educated father and a rebelliousness towards mother's aspirations, as in the case of L.K. In this case the patient is reacting to both mother's and grandmother's over-solicitousness. He uses denial extensively to deal with dependency needs.

Ambivalent Identification

Case 3. D.V. is another bright 15-year-old boy with severe PTC deficiency. In contrast to the previous case, he is doing good schoolwork and aspires to become a science teacher. He has had about 20 transfusions in the last five years for bleeding episodes but no hospitalizations have been necessary.

In exploring the patient's interests in specific subjects, it was found that there was a very close relationship between his illness

and his aspirations. He thought he would like to do work in pathology in order to "know what I'm doing". He mentioned that doctors don't always know what they are doing—he had some hair sewn into a laceration at age four causing an infection. He was critical of some doctors who did not know what "Christmas Disease" was and also of items on psychological testing materials which he considered "stupid" or "crazy".

This patient's ambitions represent a reaction formation to feelings of lack of control over the manifestations of his bleeding disorder (e.g. he wants to be "someone who takes blood"). The ambivalence towards doctors is dealt with by partial identification which in some instances has proved to be an effective solution to the conflict over continuing dependency on medical assistance.

Behavior Problems

Case 4. C.L. is a 16-year-old hemophiliac who is only in the ninth grade. He has had many traumatic bleeding episodes but is behind at school primarily because of his poor work and arguments with teachers and other students. He has been in innumerable fights, has wrecked the family car and has been in trouble with the police because of malicious destruction of property.

C. has had excessive bleeding from birth. He was diagnosed as hemophiliac at age one and had severe bleeding following tooth extraction at age six. He has had internal bleeding from the kidneys and this has been difficult to control on occasion with replacement therapy.

C. was a husky, good-looking adolescent who obviously enjoyed the interview. He described his many escapades with relish, especially getting into a physical fight with his father. There was a provocative and self-destructive quality about this. He admitted feeling depressed at times.

C.'s father is a very intelligent and well-educated, hard-driving business man who works excessive hours and makes perfectionistic demands on himself and his son. He

suffered a heart attack about five years previously but this has not slowed him down. Father carefully pointed out that the bleeders in the family are all on the mother's side, not his. C.'s mother seems to stay in the background and does not get involved in the frequent arguments.

About a year previously Mr. L. and his son consulted a psychiatrist in their home town. In the course of several interviews according to Mr. L. he was made to understand that his son's behavior was a result of his need to show that he was not weak or sick and a reaction to Mr. L.'s excessive perfectionistic demands. Since that time C.'s behavior has improved.

Sibling Loss and Exacerbation of Hemophilia

Case 5. L.C. was a 20-year-old college freshman who has been admitted to the hospital on numerous occasions since the death of his brother about three years previously. The brother was ten years older than the patient and the two were very close, often taking care of each other during bleeding episodes. The brother was a successful professional who was married and had small children.

The patient was grief-stricken at the death of the brother from hemorrhage following an automobile accident and felt that he has never gotten over it. Since his death the patient spends a great deal of time with his brother's family, and immediately prior to the current admission (for a hematoma) he had fallen down the stairs just after a visit to the dead brother's wife and children.

The numerous injuries and hospitalizations have contributed to his difficulties at school. In spite of a conscious desire to emulate his brother, he does not seem to be able to achieve comparable success at school. When seen in the hospital he was depressed and very guarded. He was reluctant to talk about his family or his feelings, and refused appointments following discharge from the hospital.

Sexual Maladjustment

Case 6. V.K. is a moderately severe hemo-

philiac who has had frequent episodes of bleeding into the joints of the lower extremity following minor trauma and wears a leg brace. He was seen for the first time at age 17 because of his refusal to enter the hospital for treatment of an effusion of the knee. At this time he made a great show of being angry about having to come into the hospital but actually was more guilty about the expenses and attention he would be receiving rather than an older brother who was also ill. He felt in some way to blame for the effusion and worried about ending up like a cousin who had a permanent disability.

Following the death of the older brother about six months later, the patient felt more guilty and wondered whether he should have died instead. The older brother's death also affected mother's behavior towards the patient in that she became increasingly protective of him especially when an operation was being considered on his knee. About the same time the patient related that he was dating a girl whom he liked a great deal but he was concerned that his love-making frequently consisted of biting and bruising the girl. By slapping his girl he provoked her to inflict bruises on him also.

On another occasion, the patient told about an incident where a young student nurse had playfully tickled his foot and he impulsively threatened to kick her in the face. In other discussions with him it became clear that his feelings about girls were exploitative rather than affectionate and his behavior in dating would consist of attempts to provoke not only the girl but her parents, e.g. by staying out late or rebelliously using his mother's car without permission. On another occasion he discussed the fact that girls were the carriers of the disease in the family. In fact, the bleeding disorders had been disseminated from his maternal grandfather by his mother and his aunts until there were now many cousins with bleeding tendencies in the family. He was concerned about the further spread of the disorder and the prospects of his own grandchildren having it.

PSYCHOSOMATICS

On one occasion in the hospital he was noisily hammering on a piece of his brace which fit into his shoe. At this point he was very close to discharge and had been told that he would have to wear his brace continually. The hammering while he was wearing the shoe resulted in hitting himself every time he hit the brace and would have been easily avoidable simply by taking the shoe off. This seemed to express his impulsive aggressiveness which he turned on to himself and which could result in self-injury.

In this young man there is evidence of guilt about dependency and sibling rivalry particularly heightened by his brother's death and the enhancement of his mother's protectiveness by that event. Along with this is a resentment and an aggressive reaction to his lack of autonomy and impaired mobility. This comes out in a kind of hypermasculine exploitation of women which has a rebellious and provocative quality and insures that it will not lead to a more mature intimate relationship. The feelings of inferiority and impaired masculinity as evidenced in his (realistic) concern about propagating the hemophilic genes prevent his achieving a comfortable relationship with girls and one can see the precursors of sexual and marital problems which are prevalent in adult hemophiliacs according to our previous study.

COMMENT

In all the cases cited, there is an attempt on the part of the adolescent to solidify his identification with the male parent either directly or through some other masculine figure (doctor, older brother, etc.). But the father is often correctly seen by the patient as rejecting his efforts to emulate him as opposed to doing "women's work". The oedipal struggle, revived in adolescence, is already compromised by persistence of unresolved feelings of helplessness and passive-masochistic gratification during many bleeding episodes. To this is sometimes added the extra burden of guilt over the death of a male sibling.

The facts of chromosomal transmission of

hemophilia offer a concrete rationale for the feelings of impaired masculinity inasmuch as the trait is maternally passed. The bleeding tendency is frequently perceived by the hemophiliac as a "curse of God" visited on him for the sins of his ancestors, thus allowing him to project guilt about his own unresolved conflicts to his mother and maternal grandfather. (The corresponding guilty feelings are found in mothers of hemophiliacs, especially over-protective ones)¹. The bleeding episodes are construed as punishment, and sometimes are actively sought by the patient himself (cases 5 and 6). The effectiveness of these mechanisms may explain the apparent lack of overt anxiety in the majority of hemophiliacs surveyed, except during acute bleeding episodes, and their relative inaccessibility to formal psychotherapy².

Thus the bleeding tendency, which is the source of much anxiety concerning bodily integrity and which aggravates unresolved sexual and aggressive conflicts, also seems to be utilized as a means of expiation of guilt feelings and avoidance of stresses in the external world. In extreme form, seen in some older hemophiliacs, there is complete reversal of husband-wife family roles and abandonment of occupation and other interests where such invalidism may not be justified on the basis of crippling. The problems encountered in adult hemophiliacs will be discussed more fully in another paper.

TREATMENT

Formal psychotherapy is neither indicated nor feasible in the vast majority of hemophiliac adolescents. As stated above, little discomfort is present in the psychic realm. Brief intervention seems effective for acute problems related to hospital management, delinquency, impairment of school performance, etc.

The most important psychotherapeutic relationship and opportunity for favorably affecting the course of development in the hemophiliac adolescent is his relationship with the internist or pediatrician who is in

overall charge of clinical management. The following principles may be useful in guiding him in his management:

1. The adolescent needs to deny evidence of weakness and dependency at the same time that he feels inadequate in his masculine endowment. A provocative show of bravado is a protest against perceived vulnerability.

2. The adolescent bleeder has an intense need for a strong accepting father or other masculine figure, including the doctor himself, to counter-balance the strong but ambivalent dependency ties to mother. The physician should not let himself get involved on mother's side in tugs of war between mother and son concerning schoolwork, choice of occupation, etc. Recommendations should be conveyed directly to the patient, not via the parents. Some counseling with fathers of bleeders is sometimes useful to convey the importance of letting the adolescent find some areas of masculine skills even if some small risk is involved.

3. The physician should recognize the

damaging psychological impact of the death of a sibling on a patient, especially if grief is inhibited. If this occurs and leads to a succession of self-induced traumatic injuries, it may be an indication for direct psychiatric intervention.

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Genius. . . has been defined as a supreme capacity for making trouble. . . It might be more fitly described as a supreme capacity for getting possessors into trouble of all kinds and keeping them therein so long as the genius remains.

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