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# Conservative management for a patient with aplastic anemia without use of blood products: case report

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## Introduction

Aplastic anemia, the failure of the pluripotent stem cell system, is characterized by pancytopenia and hypoplasia of bone marrow. The patient exhibits signs and symptoms of neutropenia and thrombocytopenia. Though the disease is rare in children, when it is seen, the peak age of incidence is between 3 and 5 years. The acquired form may occur as a toxic reaction to medications (e.g., antiinflammatory, antiepileptic, antimalarial, or antidiabetic drugs), chemotherapeutic agents, or chemicals (e.g., solvents and insecticides). It also may occur as a complication of infection, most notably hepatitis, or in association with early manifestations of leukemia. However, in half of the patients, no causative agent can be identified (Silver et al. 1987).

The most current classification of aplastic anemia consists of those patients with a severe form of the disease and those with a mild to moderate form. Severe aplasia is defined by granulocyte counts of less than  $0.5 \times 10^9/L$ , platelets less than  $20 \times 10^9/L$  and reticulocytes less than  $20 \times 10^9/L$  (Champlin 1981). A platelet count of  $20 \times 10^9/L$  is equivalent to  $20,000/mm^3$ . The moderate form of the disease is defined by at least two of the following criteria: granulocytes less than  $1.0 \times 10^9/L$ ; platelets less than  $50 \times 10^9/L$  ( $50,000/mm^3$ ); and reticulocytes less than  $60 \times 10^9/L$  (Champlin et al. 1983).

Clinical signs and symptoms include pallor, fatigue, fever and an increased tendency to bleed. Treatment varies based on whether the disease is the acquired or the congenital form, the age of the patient, and the severity of the disease. The congenital form is less common and generally is treated with blood transfusions and antibiotics, along with androgenic steroids or corticosteroid therapy, to reduce the patient's tendency to bruise and bleed. Most of these patients require continuous therapy to control the disease process (Behrman and Vaughan 1987). Treatment of the congenital or acquired forms also may consist of a bone marrow transplant from a sibling with human leukocyte antigen-compatible marrow. Some feel that this procedure yields better results in younger patients and in more severe cases (Champlin et al. 1983). In acquired cases, it can increase the survival rate by 50–70%.

Fanconi's anemia, or the congenital familial form of aplastic anemia, is believed to be inherited as an autosomal recessive trait. Though about two-thirds of these children present with other congenital anomalies (e.g.,

microcephaly, short stature, and hyperpigmentation of the skin) the pancytopenia usually is not present at birth or during infancy. Bruising secondary to the thrombocytopenia is usually the first clinical presentation; it tends to appear between 1–1/2 to 22 years of age, with an average age of 6–8 years (Behrman and Vaughan 1987).

Oral manifestations secondary to neutropenia and thrombocytopenia include purpura, spontaneous gingival bleeding, severe ulcerative stomatitis, and/or pharyngitis. Lesions may become large and necrotic, and have an accompanying foul odor. Cervical and submandibular lymphadenopathy also are common (Rose and Kaye 1983).

Dental management often requires costly and time-consuming administration of blood products before treatment. Often, parents do not want blood products administered unless absolutely necessary. The following case discusses the dental management of a pediatric patient with aplastic anemia for whom dental treatment was provided under general anesthesia on an outpatient basis without the administration of platelets.

## Case Report

A 4-year, 3-month-old white female, with a medical diagnosis of aplastic anemia, presented to Children's Medical Center of Dallas, Dallas, Texas, for dental evaluation. The mother had taken the child to a local pediatric dentist, who noted severe dental caries. The decision was made to treat the patient in the operating room because of her condition and uncooperative behavior.

The patient's past medical history was unremarkable until 10 months of age. At that time, a series of tests confirmed a diagnosis of aplastic anemia. Although the patient had many features of Fanconi's anemia, studies performed to detect Fanconi's anemia were negative. Since the time of diagnosis, she had done reasonably well, but maintained chronically low platelet counts and mild anemia. She had no problems with bleeding except frequent petechiae and easy bruising. In addition to the aplastic anemia, she also was diagnosed with developmental delay and a seizure disorder for which she was taking Tegretol® (Geigy Pharmaceuticals, Ardsley, NY).

When the patient presented for dental examination, she appeared thin, small, and microcephalic. Due to a

lack of cooperation, only a visual oral examination was performed; it revealed gross caries involving the maxillary incisors and several posterior teeth (possibly requiring the extraction of at least one tooth). Plans were discussed with the parent to perform an examination with radiographs, then complete the necessary treatment in the operating room under general anesthesia.

The preoperative physical examination showed a platelet count of 14,000/mm<sup>3</sup> (the normal value for a 4- to 5-year-old child is between 150-400,000/mm<sup>3</sup>). The hematology consultation stated that no antibiotic coverage would be needed. However, the following recommendations were made: 1) that platelets be matched and ready for transfusion should they be required; 2) that Amicar® (Lederle Laboratories, Pearl River, NY) at a dose of 100 mg/kg every 6 hr, be given starting the day before and continuing for several days after the surgery; and 3) that a room be reserved should she need to be admitted following the outpatient procedure.

The platelet count on the day of surgery was 23,000/mm<sup>3</sup>. An oral intubation was performed to avoid the possibility of bleeding. A complete examination was performed, radiographs were exposed and interpreted, and a treatment plan was formulated. Treatment using rubber dam isolation included occlusal amalgams on teeth A, J, K, L, and T; a facial amalgam on tooth M; stainless steel crowns on B and D; stainless steel crowns and pulpotomies on E, F, G, and I, and the extraction of S. Following the extraction, pressure was applied until hemostasis was achieved. The administration of platelets was not necessary.

Two hours postoperatively, the extraction site had clotted well with no oozing. The patient was discharged that same day with instructions to continue Amicar for five days to prevent clot breakdown. Postoperative instructions included pressure hemostasis with 2x2 gauzes as needed, no drinking from straws to prevent clot dislodgement and no drinking carbonated beverages for the first 24 hr. She returned to the dental clinic in one week, at which time the extraction site had healed without bleeding or problems since discharge. At the six-month recall examination, all tissues were normal and there was no recurrent decay.

## Discussion

Except for the implementation of oral hygiene procedures, and necessary therapy prior to bone marrow transplantation, it has been recommended that only emergency dental procedures be performed for aplastic anemia patients during periods of severe neutropenia. The most serious complication of dental treatment for neutropenic patients is severe, life-threatening infection. Specific dental treatment should be limited to

infection control and palliative treatment until the patient's blood values improve. Even though this patient presented with a very low platelet count, we proceeded with treatment because her medical history indicated that her values did not improve much beyond those levels. Since the patient has no siblings, future improvement via bone marrow transplant is limited.

When the platelet count is less than 50,000/mm<sup>3</sup>, prolonged bleeding following dental procedures should be considered. In spite of a platelet count of 23,000/mm<sup>3</sup>, pressure hemostasis without excessive blood loss was achieved for this patient following an extraction. The potential for bleeding should be noted when it is necessary to administer posterior superior and inferior alveolar nerve block injections because the risk of hematoma formation is high. It is felt that these injections should be avoided if possible (Rose and Kaye 1983). Hematoma formation associated with the injection of local anesthetic was avoided by using general anesthesia in this case.

Usual procedures for aplastic anemia patients before dental treatment generally include platelet transfusion and administration of epsilon amino caproic acid (EACA) or Amicar. The practitioner should, however, consider that platelet survival is decreased by fever, infection, splenomegaly, and intravascular coagulation and that platelets exhibit extreme fragility during transfusion. The decision to use platelets should be made judiciously and on an individual basis. Many aplastic anemia patients tolerate low levels of platelets without bleeding and therefore, transfusions should be reserved until levels are less than  $5 \times 10^9/L$  (5,000/mm<sup>3</sup>, Champlin 1981). The hematologist prescribed Amicar before the surgery and for five days after to prevent the breakdown of the clot. Amicar contains a synthetic monoaminocarboxylic acid with chemical characteristics similar to those of lysine. It is absorbed into the gastrointestinal tract and counteracts excessive fibrinolysis. In therapeutic dosage, it competitively inhibits the activation of plasminogen by different types of activators, such as streptokinase, urokinase, and tissue activators (Tavenner 1968).

This report demonstrates that dental care can be provided for a child with a bleeding disorder without administering platelets or other blood products on an outpatient basis.

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### **Group calls for ban on U.S. ads for snuff**

A national society of physicians has called for a ban on U.S. advertising of snuff, and has issued a warning to teenagers about the life-threatening dangers of snuff use.

The American Academy of Otolaryngology-Head and Neck Surgery cites government data showing that, since 1981, use of moist-type snuff has increased 50%, while consumption of cigarettes and eight other types of tobacco products has declined. (Snuff is flavored tobacco that is placed between the cheek and gum.)

The group recently released a survey of their physician members confirming that thousands of their patients have developed medical problems related to the use of smokeless tobacco. Of the physicians responding to the survey, 73% favored a ban on all advertising or promotion of smokeless tobacco.

"Users of snuff have been found to increase their risk of oral cancer as much as 50 times, and this disease kills 30 to 50% of its victims within five years of diagnosis," said Jerome C. Goldstein, MD, executive vice president of the Academy.

The Academy's *Through With Chew* campaign, in its third year, targets young people with the message *Snuff Out Snuff Before It Is Too Late*.