The patient with an endocardial cushion defect has many handicaps. In the incomplete type, or ostium primum, correct diagnosis was for long a problem and there seems little doubt that some of these were in the past misdiagnosed as having an atrial defect with rheumatic carditis and were slowly rescued from long-term convalescent homes and returned to normal life by correct analysis of the electrocardiogram and vector patterns. Because the general recognition of the exact anatomic features of the defect only shortly preceded the surgical era, the hazards of surgery came as a rude shock, chief being the production of complete heart block during repair of the cleft mitral valve. The risk of repair of the complete type, or AV canal, remains even now very high, in large part due to associated anomalies and to the frequent occurrence of short chordae tendineae, making the tricuspid valve or mitral valve insufficient even after repair of the clefts.

Postoperative hemolytic anemia, though rare, is another of these handicaps. The true incidence is impossible to assess, since there has been considerable reticence to publish long-term postoperative follow-up studies in this group. The first published and still the clearest account of this anemia is that of the group at Hammersmith Hospital, London.\(^1\) The patient was a 25-year-old man, with an ostium primum defect, working and supporting a family, operated on electively. Postoperatively he developed a profound and incapacitating hemolytic anemia necessitating repeated transfusions: multiple hematologic studies proved, as they have in other cases,\(^2\) that this anemia was not of an auto-immune type and was unlikely to respond to splenectomy. Since the patient was clearly worse off than he had been preoperatively, Mr. Cledland reexplored him, found a slight mitral insufficiency and adjacent to and presumably played on by the jet, a small area of exposed Teflon; he covered the latter, did nothing further to the mitral valve, and the hemolysis stopped "as if turned off by a tap."\(^3\) This rewarding experience encouraged our group to reoperate on a 5-year-old girl (case 1 in Sigler's series)\(^2\) with severe prolonged hemolysis following repair of an AV canal defect: in her case, a small nubbin of Teflon covered with platelets was found adjacent to the residual mitral valve cleft. Removal of the nubbin and repair of the residual cleft by Dr. Bahnson was followed by dramatic disappearance of the hemolytic process.

The complication of hemolytic anemia leads to yet another complication. The massive hemolysis leads to hemosiderin deposits in the
urine and secondary iron-deficiency anemia. The effects on iron metabolism were extensively analyzed by Sigler and associates 2 and are also discussed in the current issue of this journal.4 The role of iron therapy in the milder cases is of considerable interest: in Sigler's case 3 and in Sanyal's case, 4 iron plus some degree of spontaneous remission has made it possible to avoid the serious hazards of re-operation. Zinkham, 5 however, has pointed out that “Even though iron therapy might keep the anemia at a so-called tolerable level, hemolysis would continue and the amount of hemosiderin deposited in the kidneys would increase so that the patient might develop irreversible renal damage. Consequently the ultimate in therapy in this disorder might not be maintenance of adequate hemoglobin levels but prevention of renal damage.”

Efforts to understand the mechanism of hemolysis have given rise to elaborate discussions of turbulence, and the somewhat fanciful sobriquet of “Waring blender” syndrome was once used. The rare, successfully reoperated cases suggest that a combination of factors is needed, namely, a high pressure jet of blood such as may be ejected through a residual mitral cleft and an exposed or inadequately endothelialized area of Teflon, no matter how small, impinged on by this jet. If this concept is correct, the virtual absence of such anemia in the many other patients now sporting intracardiac Teflon becomes reasonable. The correctness of the term “exposed” and the role, if any, of turbulence are still debated and probably require further experimental elucidation.

The difficulties of correct diagnosis of this anemia may be compounded in severe cases by the presence of fever, splinter hemorrhages, and cardiomegaly, leading to diagnosis of endocarditis, pump fever, post-pericardiectomy syndrome, and the like: the most useful single finding appears to be the massive hemosiderin deposits in the urine although the eager and cooperative hematologist will perform many additional and relatively atraumatic studies.

This rare complication will shortly, one hopes, pass into history and be seen no more; in the meantime it has added yet another to the handicaps of the patient with an endocardial cushion defect.

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References

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The Complexity of Medicine

Medicine is a composite subject. One of its elements is an experimental science, but a large part of it obeys the very different discipline of a practical art. In consequence it has often to deal with and act upon incompletely definable situations, and to develop the faculty of practical judgement on imperfect evidence—an activity characteristically absent from an experimental science. As long, therefore, as medicine continues to be so largely an activity of a non-scientific kind, every faculty of the active, rational mind is to be desired in the practice of it.—The Collected Papers of Wilfred Trotter, F.R.S. London, Oxford University Press, 1946, p. 154.
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