Relationship between Polycythemia and Surgical Mortality in Patients Undergoing Total Correction for Tetralogy of Fallot

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Selection of the most suitable operation for patients with Fallot’s tetralogy should be based upon a knowledge of the risk involved. Results of the Blalock-Taussig operation have been thoroughly studied.1 Shunt operations can be performed with good knowledge of general risk to the patient and with assurance that the result will most often be dramatic and recovery prompt. At best, however, the result is palliative, and long-term observation of patients following this procedure suggests that it is effective for a limited time only.2,3 In contrast, surgical cardioplasty with complete correction of the defect has not been so well evaluated. Certainly, operative risk is much greater with complete correction. The theoretic desirability of plastic surgical reconstruction of the heart is obvious, however, and evaluation of these patients a year or more after surgical treatment suggests that the long-term results will be good.4

We observed that the risk of corrective operation in patients with tetralogy of Fallot who were severely cyanotic was higher than in those who were less cyanotic. Patients who had been severely cyanotic and who had benefited from a systemic-pulmonary shunt seemed to tolerate complete correction with fewer deaths than those “post-shunt” patients who were severely cyanotic at the time of complete correction.

This study was undertaken to determine the relationship between polycythemia and surgical mortality in patients undergoing total correction for tetralogy of Fallot and, further, to evaluate the possible beneficial effect of a systemic-pulmonary anastomosis prior to complete correction.

Clinical Material

Between 1954 and 1962 “corrective” operations were performed in 203 patients with tetralogy of Fallot at the Texas Children's and St. Luke's Episcopal Hospitals, Houston, Texas (table). One hundred fifty-seven patients were cyanotic (table, parts I and II), average age 11 years, and 68 had undergone previous systemic pulmonary shunts (table, part I). The over-all mortality for “total correction” in these cyanotic patients was 18 per cent. Forty-six patients had “acyanotic” or “atypical” tetralogy of Fallot and the surgical mortality was 4 per cent in this group (table, part III). The information in this paper is derived from a study of the clinical and laboratory data in the 157 patients with typical or cyanotic tetralogy of Fallot.

It is obviously difficult to categorize the severity of disease in a given patient with tetralogy of Fallot. The size of the pulmonary artery and magnitude of the right-to-left ventricular shunt are two factors that might be quantitated for this purpose. In these patients, however, it is difficult to collect a large number of patients with sufficient data to derive many conclusions concerning these factors. For this reason, patients with cyanotic tetralogy of Fallot who had a hemoglobin of 18 Gm. per cent or more were considered to be suffering from a severe form of the disease, while those with less than 18 Gm. per cent were considered as having a milder form. The age distribution of patients in the various groups was approximately the same. The “complete” surgical correction was performed by the same surgical team and post-

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operative care was provided by the same group of cardiologists. In some instances, the aortopulmonary anastomosis had been created elsewhere, and hemoglobin concentration prior to this preliminary surgery was unknown.

Patients who underwent total correction of typical tetralogy of Fallot were divided into four categories:

1. Those who had a hemoglobin concentration of less than 18 Gm. per cent prior to a preliminary shunt but who had more than 18 Gm. per cent hemoglobin concentration at the time of complete correction (table, part I, A. 1). These patients were classed as having a milder form of tetralogy at the time of preliminary shunt but who, after an initial improvement, subsequently became worse.

2. Those who had a hemoglobin concentration of 18 Gm. per cent or more prior to a preliminary shunt and who subsequently had total correction. These patients presented a severe form of the anomaly at the time of preliminary shunt and were further divided into those whose hemoglobin continued in excess of 18 Gm. per cent (table, part I, B. 2), and those whose hemoglobin returned toward normal following the shunt procedure (table, part I, B. 1).

3. Those whose hemoglobin concentration was unknown prior to preliminary shunt were divided into two groups: Those whose hemoglobin concentration was greater than 18 Gm. per cent prior to complete surgical correction (table, part I, C. 1), and those whose hemoglobin concentration was less than 18 Gm. per cent prior to complete correction (table, part I, C.2).

4. Those who had no preliminary shunt (table, part II). These patients were further divided into those who had a hemoglobin concentration of 18 Gm. per cent or more at the time of total correction (table, part II, A), and those who had a hemoglobin concentration of less than 18 Gm. per cent but who had either clinical cyanosis or laboratory evidence of right-to-left shunt (table, part II, B).

### Results

Seven of 26 patients without preliminary shunt and with a hemoglobin concentration of greater than 18 Gm. per cent died following correction (27 per cent) (table, part II, A). The risk was even higher (three deaths in five patients) in those patients who had had a preliminary shunt but whose hemoglobin concentration had returned to 18 Gm. per cent or more at the time of total correction (table, part I, B. 2). Among the 17 patients who had an elevated hemoglobin concentration at the time of complete correction, but whose hemoglobin concentration was unknown at the time of the shunt procedure, there were six deaths (35 per cent) (table, part I, C. 1). In the group of six patients whose hemoglobin was less than 18 Gm. per cent at the time of a preliminary shunt and 18 Gm. per cent or more at the time of complete correction, there was one death (17 per cent).
(table, part I, A. 1). Thus, the average mortality was 31 per cent in the 54 patients with or without preliminary shunt who had a hemoglobin concentration greater than 18 Gm. per cent at the time of total correction.

In contrast, among the nine patients whose hemoglobin concentration was greater than 18 Gm. per cent at the time of the preliminary shunt and less than 18 Gm. per cent at the time of complete correction, there were no deaths (table, Part I, B. 1). In the group of 57 cyanotic patients who had no preliminary shunt but who had a hemoglobin concentration of less than 18 Gm. per cent at the time of total correction, there were eight deaths (14 per cent) (table, part II, B). Thus, the average mortality rate following total correction for 97 patients, with or without a preliminary shunt, but with a hemoglobin concentration of less than 18 Gm. per cent, was 10 per cent.

Discussion

Other factors being equal, surgical mortality is usually directly related to the severity of the disease being treated. If the original assumption is correct, that patients with hemoglobin concentrations greater than 18 Gm. per cent have a more severe form of tetralogy of Fallot than those with lesser values, then surgical mortality should be found directly proportional to the hemoglobin concentration. This was, indeed, the case, as can be seen from the data in the table.

Patients with marked impairment (i.e., hemoglobin values of 18 Gm. per cent or more) from tetralogy of Fallot who are subjected to a shunt procedure can anticipate a much lower operative risk when total repair is performed if polycythemia has been reduced (table, part I, B. 1). Markedly polycythemic patients have been found to exhibit clotting abnormalities and have a higher incidence of postoperative hemorrhage than patients with lower hemoglobin values. Hemorrhage was the leading cause of complications and death in patients undergoing total correction, and its occurrence may be related to the degree of polycythemia. Furthermore, clinical improvement following a shunt procedure is possibly associated with a strengthening of the left heart or a conditioning of the pulmonary vascular bed in preparation for its new role when definitive repair is completed.

Since the degree of anatomic and physiologic impairment in patients with tetralogy of Fallot is difficult to quantitate, it is helpful to have a simple laboratory test to serve as a guide to these factors. Although the degree of polycythemia has been used for years as an index of the severity of disease in tetralogy, we are aware of no clinical study that has correlated this finding with surgical mortality. Our data would appear to support this connection and to indicate that the hemoglobin value may be used as a reliable index of the operative risk in patients subjected to total correction of tetralogy of Fallot.

Summary

"Complete" surgical correction of Fallot's tetralogy carries a much higher risk in patients with a hemoglobin concentration greater than 18 Gm. per cent (31 per cent mortality) than in those whose hemoglobin concentration is less than 18 Gm. per cent (10 per cent mortality).

The successful reduction of hemoglobin concentration from greater than 18 Gm. per cent to less than 18 Gm. per cent through systemic-pulmonary anastomosis greatly reduces the surgical risk associated with "complete correction." The mechanism by which this reduces surgical mortality is not known but may be related to (1) decreasing the hemorrhagic tendency associated with polycythemia and (2) conditioning of the pulmonary vascular bed and "left heart" in preparation for their new role when definitive repair has been accomplished.

The hemoglobin concentration can be used to grade the severity of Fallot's tetralogy. Such grading would provide a basis for valid appraisal of surgical mortality rates among otherwise comparable series of cases.

References

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Galen on the Movement of Blood

Galen did not clearly state whether the blood, once it passed into the pulmonary veins, was transmitted to the left ventricle. . . . Galen held that inspired air in some form or other, or some quality derived from air, was transferred from the lung through the venous artery into the left cavity of the heart by means of the diastolic active dilatation of the ventricle, and that there was a movement of waste products in the opposite direction, from left ventricle to the lung through which they were expired. "The venous artery (pulmonary vein) has no advantage of being closed since it has rather the mission of letting pass from the heart into the lungs the sooty residues which the natural heat necessarily produces in that organ (the heart) and which have no shorter means of exit. This discharge is made possible by the comparative weakness of the mitral valve."

Compounding this unfortunate assumption with another that has been a blot on his fame, Galen stated that some blood passed directly from the right ventricle into the left through invisible pores located in the interventricular septum.

Once in the cavity of the left ventricle, and only there, were blood and pneuma elaborated into the vital spirit. Through their own pulsific properties, the aorta and the arteries drew the spirituous blood from the left ventricle and distributed it throughout the body.

Galen's scheme was a decisive step toward the understanding of the movement of blood through the lungs. To be sure, it introduced the paradox of two-way traffic in the pulmonary vein, and of the selective permeability of the mitral valve for sooty wastes but not spirituous blood, both of which led William Harvey to reconsider the Galenic system. But was not this paradox the first attempt to explain the two simultaneous functions served by the movement of blood through the lungs: the acquisition of a useful substance, and the elimination of a wasteful one?—André COURNAND, M.D. Circulation of the Blood. Edited by Alfred P. Fishman, M.D., and Dickinson W. Richards, M.D. New York, Oxford University Press, 1964, p. 14.
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Circulation. 1965;32:65-68
doi: 10.1161/01.CIR.32.1.65
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1965 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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