Results of Valvulotomy for Valvular Pulmonary Stenosis with Intact Ventricular Septum

Analysis of Sixty-Nine Patients

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The analysis deals primarily with follow-up studies of patients who have had a valvulotomy for valvular pulmonic stenosis with an intact ventricular septum. In addition to observations on the subjective improvement, hemograms, and arterial oxygen saturation, the cardiac findings have been analyzed in detail. The changes in the size of the heart have been studied in relation to the growth of the patient and, wherever possible, these changes have been correlated with the postoperative right ventricular pressure. It was found that cardiac enlargement came to an end when the right ventricular pressure fell below 100 mm. Hg.

Eighty-six patients with valvular pulmonary stenosis and an intact ventricular septum had a valvulotomy performed by Dr. Alfred Blalock or one of his associates at the Johns Hopkins Hospital between November 1949 and November 1953. There have been 7 deaths in this series of 86 patients. It is of interest and of possible significance that all patients who died were cyanotic. One death occurred on the third postoperative day from an unsuspected brain abscess. Two of the other deaths occurred in patients in whom an anastomosis was performed because of the erroneous diagnosis of tetralogy of Fallot; both died at the time of the pulmonic valvulotomy. Two other patients who had had an anastomosis for tetralogy of Fallot survived a pulmonary valvulotomy; these 2 patients, however, are not included in this analysis of the operative results because the anastomotic procedure grossly altered their clinical findings. In addition, there are 8 patients in whom follow-up studies have not been adequate. Hence, this analysis of the operative results deals with the 69 patients upon whom we have postoperative follow-up studies for periods varying from 3 months to 4 years.

Subjects

Of the 69 patients, 40 were female and 29 male. The age at the time of operation ranged from 5 months to 27 years. There were 2 patients under 1 year of age; 4 between 1 and 2 years; 14 between 2 and 6 years; 24 between 6 and 12 years; 14 between 12 and 18 years; and 11 patients over 18 years of age.

Complete information on all aspects of the postoperative results was not obtained on every patient but, whenever possible, we have compared the preoperative and postoperative exercise tolerance, hemograms, and arterial saturations, and have studied the changes in the size of the heart. Finally, the changes in the size of the heart have been correlated with changes at cardiac catheterization.

Seven patients have been followed for 3 to 6 months postoperatively; 14 for 7 months to 1 year; 15 for 1 to 2 years; 11 for 2 to 3 years; and 22 for more than 3 years.

Results

Subjective Improvement. The subjective improvement in exercise tolerance following pulmonic valvulotomy has been repeatedly reported. Even though it is probably the least reliable criterion for successful operation, a careful evaluation of this feature is of some merit.

Sixty-eight of the 69 patients in this series have shown definite evidence of subjective im-

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Table 1.—Results of Preoperative and Postoperative Cardiac Catheterization in Seventeen Patients

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pt. No.</td>
<td>Age (yr.)</td>
<td>Right ventricular pressure</td>
</tr>
<tr>
<td>1</td>
<td>5</td>
<td>96/0</td>
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<tr>
<td>2</td>
<td>13</td>
<td>143/0</td>
</tr>
<tr>
<td>3</td>
<td>14</td>
<td>122/20</td>
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<td>4</td>
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<td>13</td>
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<td>6</td>
<td>7</td>
<td>166/26</td>
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<tr>
<td>7*</td>
<td>14</td>
<td>84/00</td>
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<tr>
<td>8†</td>
<td>12</td>
<td>120/40</td>
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<tr>
<td>9</td>
<td>14</td>
<td>160/3</td>
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<td>10†</td>
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<td>66/12</td>
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<td>11</td>
<td>9</td>
<td>178/0/13</td>
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<td>12</td>
<td>8</td>
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<td>13</td>
<td>15</td>
<td>148/3</td>
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<td>14</td>
<td>12</td>
<td>213/0</td>
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<td>16</td>
<td>17</td>
<td>144/26</td>
</tr>
<tr>
<td>17</td>
<td>19</td>
<td>130/2</td>
</tr>
</tbody>
</table>

* Dampened pressure.
† Patient believed to have an anomaly of pulmonary venous return.
§ N.E.—Not entered.

provement. Indeed, only 1 patient (B.I., #8 in table 1) has failed to show improvement; this patient had a 2 to 3 volume per cent step-up in the right atrium prior to and following operation, and it is believed that she has an anomaly of the pulmonary venous return in addition to pulmonary stenosis.

Prior to surgery, 19 patients, who were unable to walk 1 level city block without stopping to rest, were considered markedly incapacitated. There were 35 patients who became fatigued only after walking more than 1 block; these patients were classified as moderately incapacitated. Ten patients complained of shortness of breath on running or climbing stairs; hence, they were classified as slightly incapacitated. The remaining 5 patients were less than 2 years of age at the time of surgery, and consequently their exercise tolerance could not be evaluated. These 5 patients, however, showed excessive dyspnea on moderate exertion.

Postoperatively, 52 patients now have no limitation of activity, i.e., they are capable of participating in sports although competitive athletics are not recommended. This group includes the 5 patients whose exercise tolerance could not be evaluated prior to surgery. All 5 have now been followed for more than 1 year after operation and are considered to be leading normal, active lives. There are 15 patients who show slight limitation of activity, all of whom were incapacitated prior to operation. Two of these 15 patients have shown no demonstrable change in exercise tolerance. Both were classified as being slightly incapacitated prior to surgery, and neither is capable, at the present time, of participating in the same activities as their coevals without experiencing undue dyspnea. There remain 2 patients whose postoperative exercise tolerance cannot be evaluated, because they have been followed for only 3 months and have not yet been allowed unrestricted activity.

Cyanosis. Cyanosis, as is well known, may or may not be present in patients with valvular pulmonary stenosis with an intact ventricular septum. Prior to operation, 40 patients showed cyanosis at rest. There were 4 additional patients who were acyanotic at rest, but manifested minimal cyanosis during strenuous activity. It is worthy of note that there were 3 patients in this group who were cyanotic during early infancy in whom the cyanosis decreased as they grew older.

Postoperatively, cyanosis has been observed in only 7 patients. Three of the 7 are cyanotic at rest and 3 show minimal cyanosis with activity. Only one patient (B.I.*) has remained markedly cyanotic following valvulotomy.

Blood Cell Counts. The red blood cell counts varied with the presence or absence of cyanosis. The red blood cell counts prior to operation ranged from 3.8 million/cu. mm. to 10 million/cu. mm. There were 24 patients with counts below 5.5 million/cu. mm.; 23 patients between 5.5 and 6.5 million/cu. mm.; and 22 patients with counts over 6.5 million/cu. mm. Eight of the 69 patients had counts above 8.5 million/cu. mm. At the time of their last postoperative examination, 54 of the 69 patients had red blood cell counts below 5.5 million/cu. mm.; 14 had counts between 5.5 and 6.5 million/cu. mm. The only patient whose red blood cell count remained over 6.5 million/cu. mm. is the cyanotic patient who is thought to have an anomaly of the pulmonary venous return.
Oxygen Saturation. Oxygen saturation of the arterial blood was determined both preoperatively and postoperatively in only 26 patients. Seven had a preoperative arterial oxygen saturation above 90 per cent and showed no significant change after operation. The other 19 patients had preoperative arterial oxygen saturations that ranged from 73 to 90 per cent (fig. 1). In 16 of these patients, the postoperative arterial oxygen saturation was above 90 per cent, while in only 3 patients the arterial oxygen saturation remained below 90 per cent. In 2 of these 3 patients, the saturations were 86 and 89 per cent and, in the remaining (B.I.), the postoperative arterial saturation remained unchanged.

Heart Murmur. The cardiac findings were studied in great detail. The auscultatory findings preoperatively were fairly uniform. A harsh blowing systolic murmur along the left sternal border that was maximal in the second and third left interspace was the rule. The murmur radiated upward toward the left clavicle. In only 1 patient was the murmur maximal to the right of the sternum; this patient was thought to have an associated anomaly, possibly subaortic stenosis. A systolic thrill was palpable along the left sternal border in all but 9 patients. The second heart sound in the pulmonic area was considered to be diminished in all but 3 patients. In these 3 patients, the second sound in the pulmonic area was thought to be of slightly greater intensity than normal. Reduplication of the pulmonic second sound was never heard in this series of patients. A gallop rhythm was heard at the apex in 8 instances.

The postoperative change in the murmur was somewhat difficult to evaluate as, in many instances, the same physician did not examine the patient on both occasions. The murmur, however, was thought to be less intense in 24 instances. Furthermore, in 14 patients, a systolic thrill, which had been present preoperatively, was no longer palpable following the valvulotomy. The second sound in the pulmonic area was considered to be reduplicated in 4 patients, and of greater intensity than preoperatively in 5 other patients. Postoperatively, a short soft diastolic murmur was heard in the pulmonic area in 15 patients. The murmur was thought to represent some degree of pulmonary insufficiency. Blount and his associates have recently reported the results of open valvuloplasty of the pulmonic valve and, in 4 of the 5 patients subjected to this procedure, a diastolic murmur was heard along the left sternal border. In our series, the development of pulmonary insufficiency has not apparently altered the postoperative course of these patients.

Liver. Engorgement of the liver was noted in 24 patients prior to valvulotomy and, in 20 of these patients, pulsations at the margin of the liver were detected. Of these 20 patients, 13 were cyanotic and 7 were acyanotic. Pulsations ceased almost immediately following the valvulotomy and, in every instance, the liver decreased in size.

Edema. Edema was present in 8 patients prior to valvulotomy and was not demonstrable in any patient on follow-up examinations.

Electrocardiogram. The electrocardiographic changes following pulmonary valvulotomy have been carefully analyzed by Landtman and, therefore, are not included in this report. Landtman showed that, following surgery,
there was a positive correlation between the extent of the changes in the electrocardiogram and of the fall in the right ventricular systolic pressure. The most significant changes, indicative of improvement, occurred in those patients whose right ventricular systolic pressure fell to below 100 mm. Hg.

Heart Size. Changes in the size of the heart were studied by a comparison of the preoperative and postoperative teleoentgenograms. Both the immediate changes, those that occurred within the first 3 weeks, and the late changes, those that occurred over a period of years, were studied. X-ray films taken within the first 3 weeks following valvulotomy were available for comparison with the preoperative films in 59 instances. As was to be expected, the greatest changes occurred in the patients with the largest hearts. A review of these films showed 2 distinct changes. In the group over 18 years of age, 4 of the 5 patients with marked cardiac enlargement showed a prompt decrease in the actual size of the heart. In contrast, 4 of 5 infants, 2 years of age or under, with marked cardiac enlargement, showed an increase in the size of the heart at the time of discharge from the hospital, 3 weeks after operation. The 1 exception to this phenomenon was a 16-month-old infant in whom, in the 2 months preceding operation, the cardiothoracic ratio had increased rapidly from 60 to 67 per cent. Immediately after surgery, this patient's heart showed an abrupt decrease in size; the cardiothoracic ratio dropped to 56 per cent. This change suggested that the previous rapid increase was due to dilatation.

In an analysis of the late postoperative changes, it was necessary to take into consideration the factor of growth. Therefore, the changes in the size of the heart were studied by the method previously used by Taussig and Goldenberg, by the superposition of one x-ray film upon another, and by the calculation of the cardiothoracic ratio. There were 48 patients who had teleoentgenograms taken more than 1 year after valvulotomy. Eight of these patients were over 18 years of age and had attained their full growth. Four of these 8 patients showed a marked decrease in the size of the heart after valvulotomy; the cardiothoracic ratio decreased and thereafter remained unchanged (fig. 2). Of the remaining 4, 3 patients had hearts of normal size at the time of operation; they have shown no change on subsequent
examinations. Thus, there is but 1 patient in this group in whom the heart was enlarged prior to valvulotomy and has remained essentially unchanged in size. This patient had a marked scoliosis that rendered accurate determination of the size of the heart difficult.

There were 40 patients under 18 years of age. The analysis of the changes in the size of the heart in this younger age group showed several distinct trends. The first, and probably most favorable result, is illustrated in figure 3. After valvulotomy, the size of the heart decreased to within normal limits and thereafter the heart and chest grew together; thus the cardiothoracic ratio decreased and thereafter remained constant. There were 6 patients who showed this type of change.

In 13 instances, the heart decreased in size but remained enlarged, as shown in figure 4. In such cases, over a period of 2 or 3 years, the heart remained constant in size and the chest has grown with the child. Therefore, the cardiothoracic ratio continued to fall until the heart came to be within the limits of normality. We regard this to be a favorable sequence of events.

In 13 patients, the changes were less favorable. In these patients, the heart remained the same size or decreased slightly in size during the first 6 months after operation; thereafter, it continued to grow with the chest. In such instances, although the cardiothoracic ratio remained constant, the heart remained enlarged (fig. 5). The lack of progressive cardiac enlargement indicates an improvement, but it is not a satisfactory result.

Finally, in this group of 40 patients, there were 8 in whom the heart was normal in size at the time of operation and has remained normal in size on subsequent postoperative examinations (fig. 6). These patients were operated upon because of dyspnea on exertion, cyanosis, or high right ventricular pressure; although the right ventricular pressure was reduced, there was no appreciable change in the size of the heart. Obviously, such patients have a constant cardiothoracic ratio.

At present, 3 children appear to show evidence of progressive cardiac enlargement. Each of these children showed a decrease in

Fig. 4. A slight decrease in the size of the heart following valvulotomy with a subsequent fall in the cardiothoracic ratio due to growth of the chest without proportional increase in size of the heart.

the size of the heart after valvulotomy; nevertheless, the heart remained enlarged. The cardiothoracic ratio, however, is again increasing. Needless to say, such patients must

Fig. 5. The heart is enlarged but cardiothoracic ratio has remained constant following valvulotomy (patient no. 13, table 1).
be kept under close observation and may require further surgery.*

The comparison of pulmonary vascularity, as visualized in the x-ray films, showed no significant change. In 9 patients, there appeared to be an increase in the size of the main pulmonary artery after the valvulotomy. In 7 of these 9 patients, it was believed that the apparent increase was in reality due to a decrease in the size of the right ventricular outflow tract.

Cardiac Catheterization. Cardiac catheterization was not a routine procedure either before or after operation. Although 48 patients had a cardiac catheterization prior to surgery, only 17 patients have had both a preoperative and postoperative cardiac catheterization. Furthermore, as is shown in table 1, cardiac catheterizations were performed at varying times after operation; 1 as early as 1 month, and 3 approximately 2 years following surgery.† It is of interest that in 2 of the 3 patients in this series (nos. 16, 17), who were catheterized approximately 2 years after surgery, the right ventricular pressure has fallen to nearly normal levels. In 4 instances, the right ventricular pressure remained over 100 mm. Hg and in one it was 90 mm. Hg. In this group the longest time interval was 13 months. Whether the right ventricular pressure continues to fall or ever returns to normal limits must await further investigation. In this connection, it is worthy of note that Soulie and his associates found the most significant drop to occur in those individuals whose initial right ventricular systolic pressure was above 150 mm. Hg. These authors also concluded, from physiologic data obtained on patients who had 2 postoperative cardiac catheterizations, that the greatest fall in pressure occurred in the immediate postoperative period. We have no information on this point, as none of the patients in this series have had a second postoperative catheterization.

The correlation of the right ventricular pressure and the change in the size of the heart is of interest. Our observations indicate that the size of the heart is not solely determined by the height of the right ventricular pressure. The degree of hypertrophy, if concentric in type, may be considerable without demonstrable evidence of cardiac enlargement by x-ray or fluoroscopic examination. The compensation of the patient is also a significant factor in determination of right ventricular pressure. For example, patient no. 10 was in severe cardiac failure preoperatively with marked hepatomegaly and pedal edema at the time of her first cardiac catheterization; her right ventricular pressure was only 66/12 mm. Hg. Eight months after valvulotomy, the pressure was essentially the same, but the heart had decreased from a cardiothoracic ratio of 66 per cent to 45 per cent (fig. 2), and there was no clinical evidence of cardiac decompensation.

As was to be expected, in those individuals in whom there was no cardiac enlargement, a decrease in right ventricular pressure did not change the size of the heart. In those patients with cardiac enlargement, the decrease in the size of the heart, after valvulotomy, was not related to the magnitude of fall in pressure, but to the level to which the pressure fell.

* One patient has subsequently shown a decrease in the size of the heart.
† We are indebted to Dr. R. J. Bing, Dr. H. T. Babnson, and their associates for these studies.
Furthermore, in patients in whom the right ventricular pressure returned nearly to normal, the heart returned to within normal size within the first postoperative year and, thereafter, the heart grew normally with the growth of the chest.

It was of great interest that, in all patients in whom the right ventricular pressure was reduced below 100 mm. Hg, even though the pressure remained above 60 mm. Hg, the heart showed a definite decrease in size and, thereafter, remained constant while the child grew. Therefore, over a period of years, the cardiothoracic ratio returned to normal.

In contrast to this finding, the 3 patients (nos. 4, 5, 13) in whom the postoperative pressure remained above 100 mm. Hg have been followed for more than 3 years and their hearts and chests have grown proportionately. Consequently, in these 3 patients, the cardiothoracic ratio has remained constant. None have shown evidence of progressive enlargement but, nevertheless, enlargement persists.

These observations indicate that a pressure of approximately 100 mm. Hg seems to be the critical level as far as the size of the heart is concerned. If the right ventricular pressure is reduced to less than 100 mm. Hg, the heart decreased in size or remained stationary while the chest grew.

**Summary**

Transventricular pulmonary valvulotomy was performed at the Johns Hopkins Hospital on 86 patients with pulmonary valvular stenosis and an intact ventricular septum. There were 7 deaths, a mortality rate of 8.1 per cent, which includes 1 patient who died of a brain abscess and 2 patients who had previously had anastomotic procedures for an erroneous diagnosis of a tetralogy of Fallot. Of the 79 surviving patients, 8 have not had adequate follow up study. Among the remaining 71 patients, there are 2 patients who had previously been operated upon for a tetralogy of Fallot. The analysis, therefore, deals primarily with 69 patients who have been followed from 3 months to 4 years postoperatively, all of whom had been operated upon prior to November 1953.

Subjective improvement was noted in all but 2 patients. The comparison of the hemograms and arterial oxygen saturations shows improvement in all patients except 1, in whom an associated anomaly of the pulmonary venous return is suspected.

The analysis of the postoperative physical findings showed the murmur to be less intense in 24 patients and the disappearance of a systolic thrill in 14 of these patients. In 15 patients, a diastolic murmur was heard in the pulmonic area. Enlargement of the liver was not noted in any patient after valvulotomy and, in every instance in which pulsations at the liver edge were present preoperatively, the pulsations ceased almost immediately following operation.

Changes in the size of the heart, both in the immediate and late postoperative periods, were studied. The results showed that if the heart was not enlarged preoperatively, there was no change in the size of the heart, whereas, when the heart was enlarged, several distinct types of change were noted. In patients under 2 years of age, there was an actual increase in the size of the heart in the immediate postoperative period. Later examinations, however, showed the heart to have remained stationary while the chest grew; thus, ultimately, there was decrease in the cardiothoracic ratio. In patients over 18 years of age, the heart decreased steadily in size during the first few postoperative months. In some of the younger patients, the size of the heart decreased to within normal limits and then grew normally with the growth of the chest; in others, the heart remained the same size while the chest grew, so that the cardiothoracic ratio decreased until the heart returned to normal size; in still others, only progressive enlargement ceased; the heart and chest grew together and the cardiothoracic ratio remained unchanged.

Analysis of the cardiac catheterization showed a correlation between the postoperative pressure and the change in the size of the heart. Those patients in whom the pressure was reduced to below 100 mm. Hg, the size of the heart returned to normal. These patients were considered to have obtained an excellent result from surgery.
**SUMMARIO IN INTERLINGUA**

In le curso de quatro annos, valvulotomia pulmonar transventricolar esseva execute al hospital Johns Hopkins in 86 patientes con stenosis pulmono-valvular e intacte septo ventricular. Occurreva 7 mortes. Isto esseva un mortalitate de 8,1 pro cento, includente un patiente qui moriva ab un absorso cerebral e 2 patientes previemente subjiciate a interventiones anastomotic post erronee diagnoses de tetralogia de Fallot. Le gruppo de 79 supervientes include 8 con inadequate observationes postoperatori. Duo del remanente 71 patientes habeva previemente essite subjiciate a interventiones pro tetralogia de Fallot. Per consequente le analyse se concerne primariamente con un gruppo de 69 patientes. In lor casos le observationes post-operatori occupava periodos de inter 3 menses e 4 annos.

Melioration subjectiva esseva reportate in omne cases con 2 exceptiones. Le comparation del hemogrammas e del saturationes oxygenic revela melioration in omne patientes con un exception. Isto concerne un patiente in qui iha un suspicite anomalia del retorno pulmonovenose.

Le analyse del postoperatori observationes physic monstrava murmures de intensitate reducitur in 24 patientes. In 14 de iste casos, etiam le disparition de un fremito systolic esseva constatate. In 15 patientes un murmure diastolic esseva audibile in le area pulmonic. Turgescentia del hepate non esseva notate in ulle patiente post le execution de valvulotomia, e in omne cases in que pulsationes del margine hepatic esseva presente ante le operation, iste pulsationes cessava quasi immediatamente post le operation.

Alterationes in le dimensiones cardiac esseva studiate tanto immediatamente como etiam plus longemente post le operation. Le resultatos monstrava que si le corde non esseva allargate ante le operation, il habeva nulle alterationes del dimensiones cardiac. In le cases in que le corde habeva essite allargate ante le operation plure typos de alteration esseva distinguibile. In patientes de etates de minus que 2 annos, il habeva de factor un augmento del dimensiones del corde in le periodo immediatamente post le operation. Examines executate plus tarde monstrava, nonobstante, que le dimensiones del corde habeva remanite constante durante que le thorace se expandeva. Ultimemente il habeva consequentemente un reduction del proportion cardio-thoracic. In patientes de plus que 18 annos, le dimensiones del corde decresceva uniformemente durante le prime menses post le operation. In certes inter le plus juvane patientes, le dimensiones del corde decresceva usque a valores intra le limites normal, e postea illos cresceva normalmente con le crescentia del thorace. In altere casos le corde non alterava su dimensiones durante le crescentia del thorace, de maniera que le proportion cardio-thoracic decresceva usque le dimensiones del corde esseva normal. In ancora altere casos, solmente le allargamento progressivo se arrestava. Le corde e le thorace cresceva parallelamente e le proportion cardio-thoracic remaneva stabile.

Le analyse de catheterisation cardiac reveleva un correlation inter le pression postoperatori e le alterariones del dimensiones cardiac. In le patientes in qui le pression se reducive usque a infra 100 mm Hg, le dimensiones del corde retornava a valores normal. Iste patientes, in nostre opinion, habeva derivate excellenti beneficiab ab le intervention chirurgic.

**REFERENCES**

2. **Sellors, T. H.:** Surgery of pulmonary stenosis; a case in which the pulmonary valve was successfully divided. Lancet, 1: 988, 1948.
5. **Taussig, H. B., and Goldenberg, M.:** Roentgenologic studies in the size of the heart in childhood; 3 different types of teleoentgenographic changes which occur in acute rheumatic fever. Am. Heart J. 21: 440, 1941.
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