Venous Hum of the Cruveilhier-Baumgarten Syndrome

Response to the Valsalva Maneuver

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The response of the venous hum of the Cruveilhier-Baumgarten syndrome (CBS) to respiration, the cardiac cycle, and changes in posture is not predictable. The Valsalva maneuver may result in an increase in intensity of the hum rather than a decrease. The Cruveilhier-Baumgarten venous hum (CBS) is virtually diagnostic of portal vein hypertension. It may be mistaken for obscure cardiac murmurs. The Cruveilhier-Baumgarten venous hum, unlike the cervical venous hum (CVH), is never present in normal people.

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The Cruveilhier-Baumgarten syndrome (CBS) includes cirrhosis of the liver, portal venous hypertension, systemic collateral venous circulation, caput medusa, venous hum, thrill, and splenomegaly. The response of the venous hum of the CBS to respiration, the cardiac cycle, and change of posture, unlike the cervical venous hum (CVH), is unpredictable. The response of the venous hum of the CBS to the Valsalva maneuver has rarely, if ever, been examined. The CVH is obliterated by this maneuver. I have observed in six patients with CBS an increase rather than a decrease in intensity of the venous hum with the Valsalva maneuver. Two illustrative cases are presented.

REPORT OF CASES

Case 1.—A 46-year-old man with biopsy-proven Laennec's cirrhosis had a splenorenal shunt performed for bleeding esophageal varices in 1964. On follow-up admission to the hospital in 1969 he was found to have a large periumbilical caput medusa (Fig 1) with a thrill, a very loud, roaring continuous venous hum, which radiated to the lower portion of the sternum and was increased by the Valsalva maneuver (Fig 2). The hum was obliterated by pressure to the caput medusa. Blood samples after an oral glucose load were drawn simultaneously from an antecubital vein and a vein of the caput medusa (Table), proving a connection between portal and systemic circulation.

The hepatic vein wedge pressure was elevated at 22 mm Hg.

Case 2.—A 65-year-old man with biopsy-proven Laennec's cirrhosis was admitted to the hospital in 1970 with increasing abdominal girth. He had a ballotable liver and splenomegaly. There were no superficial abdominal varicosities. A continuous sighing venous hum was heard from just below the xiphoid of the sternum to the fourth and fifth left intercostal spaces adjacent to the sternal border. The hum was made louder with the Valsalva maneuver and was obliterated by light pressure applied to the left of the xiphoid process. With the hum obliterated by pressure, a diastolic murmur indistinguishable from aortic regurgitation was heard along the left sternal border.

The hepatic vein wedge pressure was elevated at 23 mm Hg.

COMMENT

The venous hum of the CBS results from the flow of blood from the higher pressure portal venous system to the lower pressure systemic circulation. The flow reaches the thoracicacoabdominal veins through the paraumbilical veins and the terminal portion of the umbilical vein. The increase in flow results in tortuous varicosities. Turbulence and a resultant venous hum may occur at abrupt angulations or succu-
Results of a Three-Hour Oral Glucose Tolerance Test

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<tr>
<th>Time</th>
<th>Adominal Vein</th>
<th>Formal Vein</th>
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<tr>
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<tr>
<td>30 min</td>
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<td>190</td>
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<td>3 hr</td>
<td>108</td>
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*Blood samples were drawn simultaneously from an antecubital vein and a vein of the caput medusa. The values for the plasma glucose are in milligrams per 100 ml.

Fig 1.—Infrared photograph of the caput medusa superior to the umbilicus.

Fig 2.—Recording of Cruveilhier-Baumgarten hum. Arrow indicates onset of the Valsalva maneuver. ECG was recorded simultaneously. Recording was made with a crystal microphone and a multichannel recorder.

The changes in venous pressure that occur with inspiration, diastole, and posture may not affect the hum of the CBS as the pressure in the collateral circulation is higher than in the systemic. Also, the mechanism of production of the hum of the Cruveilhier-Baumgarten syndrome may vary. The flow of blood in the collateral circulation may be away from, rather than toward, the heart. The CVH is louder during inspiration and cardiac diastole and with the patient in the upright position. The Valsalva maneuver obstructs venous return and the cervical venous hum ceases.3

Increased intensity of the CBH with the Valsalva maneuver is unexplained. Muscular contraction may constrict a penetrating vein and produce turbulence or the abdominal muscle contraction may completely obstruct flow in one vein with a secondary increase in flow in others. Constriction of the vein by muscular contraction may obliterate the hum.5

A superficial abdominal hum, with or without obvious varicosities, is virtually diagnostic of portal vein hypertension. The CVH and the CBH may be transmitted to the precordium and be mistaken for obscure cardiac murmurs. This confusion is resolved by compressing the vein and obliterating the hum.

The hum of the CBS, if heard, is an important clue to the presence of portal vein hypertension and provides a clue to the origin of ascites before other procedures are done.

Anne S. Erwin prepared the manuscript.

References