hemorrhagic pulmonary infarction. They appear during an acute phase of the disease and resolve within a few days. Proximal opacities near the hila correspond to pulmonary artery aneurysms. Hughes-Stovin syndrome, which affects young male subjects, has been defined as the association of venous thrombosis and pulmonary artery aneurysms. Hemoptysis, fever, and intracranial hypertension or optic neuritis have been mentioned in connection with it. The histologic aspect of the syndrome consists of pulmonary artery vasculitis closely resembling the vasculitis in BD. It is felt now that Hughes-Stovin syndrome is a forme fruste or unrecognized BD. The characteristic pathologic feature is a leukocytoclastic vasculitis that is felt to be secondary to immune complex deposition.\textsuperscript{4,5,7} IgG, C\textsubscript{1}, and C\textsubscript{3} have been reported to be present in small pulmonary veins and alveolar septal capillaries.\textsuperscript{7} Pulmonary arteries of all sizes can be involved leading to aneurysms and bronchial erosion secondary to these aneurysms. It has been suggested that pulmonary arterial hypertension promotes the creation of pulmonary arterial aneurysms. In our case, pulmonary artery pressure was normal. The pulmonary aneurysms in BD probably develop as the inflammatory process gradually destroys the elastic elements of the media, with subsequent dilatation of the vessel wall.

Various criteria have been formulated in order to make a diagnosis of the disease and consist of recurrent aphthous ulcerations (either oral or genital) and two of the following criteria: uveitis, synovitis, cutaneous vasculitis, or meningoencephalitis.\textsuperscript{4,5,7} Studies from different parts of the world have shown several cases with incomplete BD and patients do not fulfill the criteria for diagnosis.\textsuperscript{1,8} In our patient, the clinical manifestations and pathologic findings eliminated other systemic vasculitis syndromes such as polyarteritis nodosa, Takayasu’s arteritis, and drug-induced vasculitis which were considered in the differential diagnosis. The possibility of infectious pneumonia was excluded by special stains, culture and serologic studies for microorganisms.

Our opinion is that some cases of BD involve the lung and may manifest as pulmonary aneurysms with hemoptysis. Therefore, we believe that this patient had BD, and pulmonary aneurysm could be considered as the first appearance of the disease.

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REFERENCES


Acute Pulmonary Hypertensive Crisis in a Patient with Primary Pulmonary Hypertension Treated by Both Epoprostenol (Prostacyclin) and Nitroprusside*

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A 19-year-old girl was diagnosed as having primary pulmonary hypertension that was confirmed by right heart catheterization. Acute right heart failure was associated with syncope. Stabilization, while not achieved with intravenous epoprostenol (Prostacyclin) alone, was achieved with intravenous prostacyclin and nitroprusside.

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We report the case of a young girl with primary pulmonary hypertension in whom a combination of intravenous nitrate together with high-dose intravenous epoprostenol (Prostacyclin)\textsuperscript{*} may have arrested a pattern of recurrent syncope associated with pulmonary hypertensive crises and right ventricular failure.

CASE REPORT

A 19-year-old girl was first referred to us for heart-lung transplant in 1988. She had complained of dyspnea for 18 months and syncope for two months. At this time, the jugular venous pressure was raised and a loud P2, a pulmonary ejection click, a right ventricular third heart sound, and a tricuspid regurgitant murmur were present. The proximal pulmonary arteries were enlarged and there was “pruning” of the peripheral vessels. Pulmonary angiography failed to show any thrombotic-embolic changes. Treatment was started with oral warfarin and twice daily administration of 20 mg of slow-release tablets of nifedipine. A month after treatment was commenced, a 12-minute walk showed her exercise tolerance to be 720 m. Over the next month, because of two syncope attacks, she required admission to hospital. The dose of nifedipine was increased to 20 mg, three times a day. There were two unexplained episodes of atrial tachycardia, and further syncope attacks prompted return to our hospital.

Right heart catheterization showed cardiac output of 2.7 L/min (cardiac index [CI], 1.7 L/min\textsuperscript{m2}), mean pulmonary artery pressure of 69 mm Hg (115/52 mm Hg), mean systemic arterial pressure of 54 mm Hg (95/41 mm Hg) at a heart rate of 94 beats per minute, and right atrial pressure of 5 mm Hg. A short-term trial of

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Following Second Collapse

Incremental doses of intravenous PGI₂ up to a maximum of 6 ng/kg/min led to a rise in cardiac output to 3 L/min (CI, 1.8 L/min/m²) with no change in mean pulmonary artery pressure or mean systemic arterial pressure. The heart rate remained at 94 beats per minute and the right atrial pressure was 7 mm Hg. Twenty minutes after stopping the PGI₂ infusion, the patient complained of nausea and became unconscious, but there was no change in heart rate (98 L/min). Epoprostenol (Prostacyclin) therapy by peripheral line was restarted (5 ng/kg/min) with improvement; however, a further syncopal episode occurred. A Swan Ganz catheter was inserted, and the rate of infusion of epoprostenol was increased to 12 ng/kg/min. During a subsequent third syncopal episode, we recorded the mean pulmonary artery pressure, which rose to 92 mm Hg, and this was associated with a bradycardia of 50 beats per minute and a mean systemic arterial pressure of 60 mm Hg. The right atrial pressure, just prior to her syncope, had risen to 13 mm Hg (Fig 1).

A nitroprusside infusion was started at the time of the syncope at 10 µg/min increasing to 15 µg/min. After 48 hours, the nitroprusside was replaced by an isosorbide infusion at a rate of 15 µg/min. There were no further syncopal episodes over the next 48 hours, during which the cardiac output rose to 5.5 L/min (CI, 3.5 L/min/m²), the mean pulmonary artery pressure fell to 60 mm Hg, and the right atrial pressure fell to 2 mm Hg. The isosorbide dosage was slowly reduced and stopped, when the cardiac output was 5.6 L/min and the mean pulmonary artery pressure was 67 mm Hg. While receiving 12 ng/kg/min of PGI₂ alone, the patient returned to the general ward and had no further syncopal attacks. Twelve months later, at home, with an intravenous long-term infusion of PGI₂ she is well and her 12-minute walking distance is 740 m.

DISCUSSION

The use of nitroprusside and other intravenous nitrates as an adjunct to epoprostenol (Prostacyclin) in acute crises in primary pulmonary hypertension has not been previously reported (to our knowledge). Syncopé in this patient was associated with an acute rise in pulmonary artery pressure and a fall in cardiac output suggesting a rise in pulmonary vascular resistance, which in turn was associated with failure of right ventricular function as shown by the rise in right atrial pressure. The combined effect of PGI₂ and nitroprusside appears to have controlled this tendency. Whether the continued clinical improvement observed without nitroprusside and later without even isosorbide was the result of reversing right ventricular ischemia is uncertain. The nitrovasodilators increase intracellular levels of cyclic GMP within vascular smooth muscle, so causing relaxation. By contrast, epoprostenol causes smooth muscle relaxation by increasing cyclic adenosine monophosphate (AMP) in smooth muscle. The two types of vasodilator given together may therefore have an additive effect. Syncopal attacks in patients with primary pulmonary hypertension indicate a poor prognosis. This feature together with a low CI and low pulmonary artery oxygen saturation carried a grave prognosis for this patient. Our patient demonstrates that acute right ventricular dysfunction associated with a rise in pulmonary vascular resistance drops the cardiac output sufficiently to cause syncope. Sudden death is the usual manner of demise in primary pulmonary hypertension. This case illustrates one possible mechanism and indicates that acute intervention with adequate amounts of intravenous vasodilator can be successful. We do not, however, advocate the use of nitroprusside generally in such patients as it is our impression that response to nitroprusside and other vasodilators can be highly individual.

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