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*Images in Congenital Heart Disease*

# Postoperative "white-out" from superior caval venous obstruction

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An eleven-day-old baby with Goldenhar syndrome, or occuloauriculovertebral dysplasia, underwent correction of mixed supracardiac and cardiac totally anomalous pulmonary venous connection using a pericardial baffle to close the sinus venosus interatrial communications and direct the right pulmonary veins into the left atrium. Post-operatively, he had prolonged and repeated drainage of large volumes of fluid bilaterally through the pleural tubes. This became chylous with enteral feeding. In the absence of drainage tubes, his chest radiograph showed a complete "white-out" (Fig. 1), with general haziness of the lung fields due to bilateral pleural effusions and pulmonary lymphangiectasia. The heart remained remarkably normal in size, and there were no signs of cardiac failure. Pulmonary venous obstruction was excluded by echocardiography and cardiac catheterization. The right ventricular systolic pressure was normal at 26 mm Hg, and ventricular function was normal.

Prominent distended veins developed on the chest wall (Fig. 2), and flow within them was noted to run inferiorly. At this time, an intercostal drainage catheter was in place. Venography performed in both arms demonstrated complete obstruction of the superior caval vein, the brachiocephalic vein, and the right subclavian vein. Numerous collateral veins from the chest wall and paravertrebral system were seen draining to the to the inferior caval vein (Fig. 3).

Four weeks after the original operation, surgical relief of the obstruction was performed, with thrombectomy of the brachiocephalic and superior caval veins, and augmentation of the junction of the right atrium with the superior caval vein using a pericardial patch extending into both branches. The patient survived for twelve months.

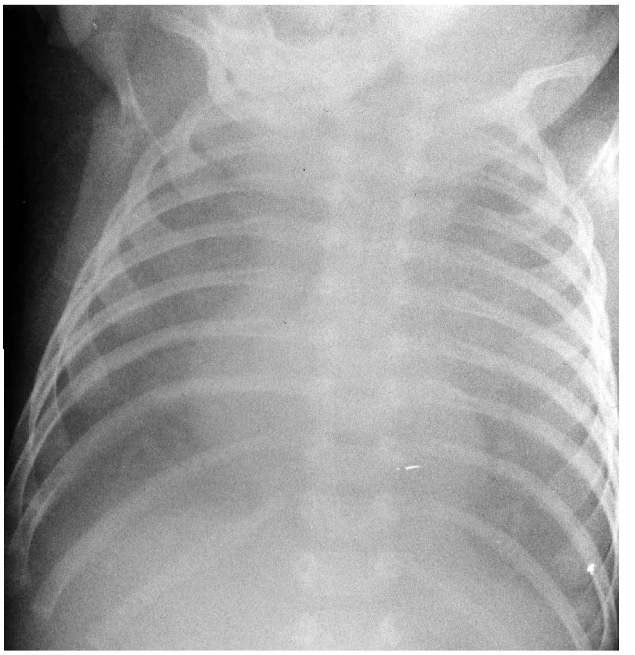

**Figure 1.**
*Postoperative chest radiograph showing general haziness of lung fields related to bilateral pleural effusions and pulmonary lymphangiectasia. The size of the heart remains normal*

The mechanism of chylous pleural effusions and pulmonary lymphangiectasia in infants with blockage of the superior caval vein is thought to be obstruction of thoracic lymph flow into the venous system.[1]

## Reference

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**Figure 2.**
*Distended veins are seen on the abdominal and chest walls draining towards the inferior caval vein. An intercostal catheter is in place to drain the recurrent large pleural effusions.*

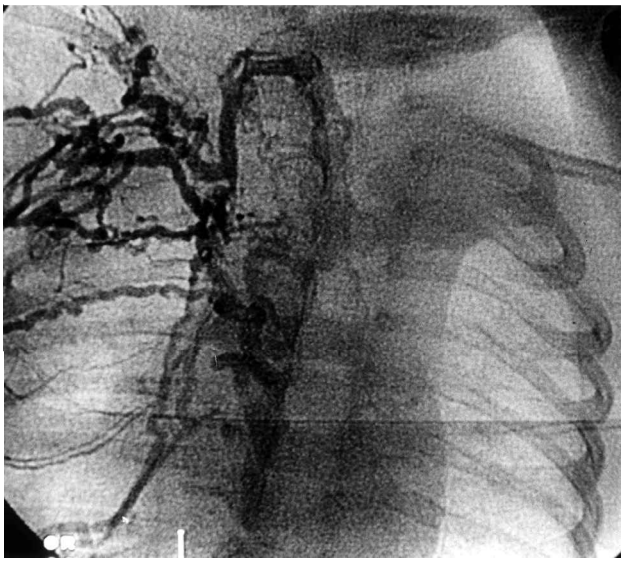

**Figure 3.**
*Right arm venogram: There is obstruction of superior caval, brachiocephalic, and right subclavian veins, with numerous distended collateral veins between the paravertebral system and the chest wall.*

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