of the tumour, and confirm that there were no intraabdominal metastases. Subdiaphragmatic venous congestion prevented more than minimal preliminary dissection.

The incision was extended into a median sternotomy and the intravascular tumour was found to extend into the lower right atrium. Cardiopulmonary bypass was established through the right atrial appendage and the aortic root. The patient was cooled to 20°C and the circulation slowed. This allowed mobilisation of the tumour mass and subsequent division of the renal pedicle at the hilum, giving access to the inferior vena cava below a grossly enlarged and engorged liver.

Circulatory arrest was then induced, the superior vena cava was snared, and the inferior cavoatrial junction opened. The combined subhepatic and suprahepatic approach allowed complete removal of the tumour thrombus from the inferior vena cava and hepatic veins. This was accompanied by visible decongestion of the liver. The venotomy and atriotomy were closed and the patient rewarmed to 37°C before discontinuing bypass without difficulty. The chest and abdomen were closed with routine drainage.

Her postoperative recovery was uneventful. She was extubated on the fifth day after operation, at which time she was fully conscious (Glasgow coma scale 14). Histology showed a nephroblastoma with a 'favourable' histological appearance, and confirmed tumour fragments within the intravascular thrombus. Chemotherapy was delayed for two weeks until liver function tests had improved, and the patient is now receiving a chemotherapy regimen modified from the United Kingdom Children's Cancer Study Group protocol for stage II nephroblastoma with favourable histology, and is tolerating this treatment well. Satisfactory control of her phenylketonuria was eventually regained 11 weeks after operation.

## Discussion

Intravascular extension of hepatocellular, adrenal, or renal carcinoma is a well recognised cause of the Budd-Chiari syndrome[2] but rarely presents with encephalopathy. Intra-atrial extension may occur in up to 0·7% of cases of Wilms' tumour[3]; we have, however, found only one previous report of Budd-Chiari syndrome in this condition.[4] We believe this to be the first reported case of fulminant hepatic failure caused by hepatic venous obstruction in a patient with a Wilms' tumour.

Intraventricular monitoring of intracranial pressure has been questioned in the management of hepatic encephalopathy in view of the risk of intracranial haemorrhage.[5] In this case vigorous treatment was required to maintain cerebral perfusion pressure before operation, and the therapeutic option of withdrawal of cerebrospinal fluid was invaluable in stabilising the patient. There were no complications.

Fulminant hepatic failure in children has a mortality of almost 70%, but the hepatic and neurological prognosis for survivors is good.[6] This case illustrates a rare but reversible cause of fulminant hepatic failure and the use of circulatory arrest in facilitating removal of an otherwise inoperable tumour. The successful outcome resulted from a multidisciplinary approach to management and, despite the patient's critical condition, early operative decompression after stabilisation was both feasible and lifesaving.

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# Hydrocephalus, bronchiectasis, and ciliary aplasia

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## Abstract

**A girl presented in the neonatal period with hydrocephalus, bronchiectasis, and ciliary aplasia. A common defect both in respiratory tract cilia and in ventricular ependyma cilia may explain the association of the two diseases.**

Hydrocephalus has recently been described in a child in association with primary ciliary dyskinesia.[1] Although hydrocephalus is not a constant feature of the disease, the association may be of interest in suggesting a functional role for the cilia lining the ventricular ependyma of the brain and spinal cord. In this paper we report a particular form of primary ciliary dyskinesia (ciliary aplasia) in a girl with bronchiectasis who developed hydrocephalus in the neonatal period.

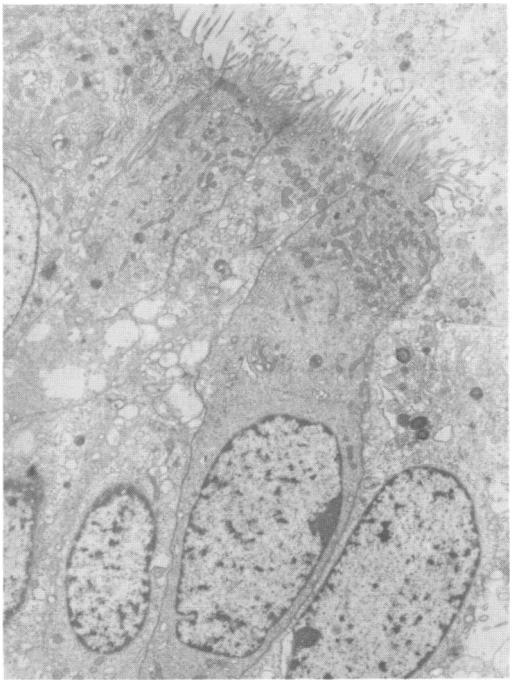
## Case report

A girl, the first child of unrelated parents, was spontaneously delivered at full term after a normal pregnancy. At the age of 3 weeks an increased cranial circumference was noted. Computed tomography showed triventricular hydrocephalus caused by stenosis of the aqueduct of Sylvius. A ventriculoatrial shunt

was inserted at the age of 1 month. She tolerated the shunt well and her subsequent neurological development was normal. Persistent respiratory symptoms comprising cough with bronchial secretions and rhinorrhoea developed during the first days of life causing recurrent infections of the upper and lower respiratory tract.

When the girl was admitted to our centre, at the age of 7·8 years, she had a chronic cough with purulent sputum and rhinorrhoea. A chest radiograph showed bilateral overinflation, increased linear shadows, and atelectasis of the middle lobe. A bronchogram confirmed the presence of diffuse cylindrical bronchiectasis and stenosis of the middle lobe bronchi. Radiographs of the sinuses showed mucosal thickening and diffuse opacification of the antra. There was moderate hearing loss for higher frequencies, and impedence audiometry was abnormal. Mucociliary clearance assessed with the saccharin test gave doubtful results because of the poor cooperation by the patient.[2] There was no evidence of any dysmorphism.

Samples from nasal and bronchial mucosa showed no particular changes in the respiratory epithelium on light microscopy. No squamous metaplastic epithelium or evidence of inflammation were seen. Both bronchial and nasal epithelium contained (among the goblet cells) numerous surface lining cells that were characteristically columnar in shape, with long, thin, sinous projections. On electron microscopy, no cells with kinocilia, basal bodies, or centrioles were seen. The goblet cells in both nasal and bronchial epithelium looked normal, and were distended by numerous and large secretory droplets. In all samples, ciliated cells were replaced by abnormal, respiratory, cilia lacking cells (figure). These abnormal cells had long projections without internal axonemal structures on their free surface. Apart from these they had the same morphological features as the normal ciliated cells. They had nuclei in their bases, numerous apical mitochondria, few lysosomes, a moderate amount of endoplasmic reticulum, and a well developed Golgi apparatus located above the apical side of the nucleus. Centrioles, basal bodies, accessory fibres, and any other structure related to centriolar precursor material were not seen within the cytoplasm of the abnormal respiratory cilia lacking cells. Compared with normal ciliated cells, they seemed to have no glycocaliceal bodies in their luminal surfaces whereas, in normal respiratory epithelia, all the goblet cells had glycocaliceal bodies in the glycocalix around the microvilli.



*Electron micrograph of bronchial mucosa. Tall columnar cells without basal bodies, centrioles, and kinocilia are present instead of normal ciliated cells. Slide stained with uranyl acetate-lead citrate, magnification ×8000.*

## Discussion

Ultrastructural examination of multiple samples from nasal and bronchial mucosa in a girl with a history of hydrocephalus and a clinical picture compatible with primary ciliary dyskinesia showed morphological features consistent with ciliary aplasia.[3] It is well known from studies carried out on animal mutants that primary defects of genes coding for axonemal polypeptides, when present, may be found in virtually all somatic cells of the same animal including the epithelium of trachea, oviduct, and ependyma.[4 5] The association between respiratory problems and the development of hydrocephalus, which has been described in a particular strain of mice, could arise as a consequence of defective ciliary axonemes, suggesting a functional role for the cilia that line the ventricular ependyma of the brain and spinal cord during embryonic development and during the normal flow of cerebrospinal fluid.[4] For this reason, in our patient it is conceivable that the absence of kinocilia in the respiratory epithelium may also have occurred in the ependymal epithelium. Although the presence of hydrocephalus was noted in a previous series of patients with Kartagener's syndrome,[6] hydrocephalus is not a constant feature in primary ciliary dyskinesia. It has been suggested that ependymal ciliary defects as described in primary ciliary dyskinesia, may be only a contributory embryological factor in the pathogenesis of particular forms of hydrocephalus. It is likely that several of the primary structural defects that interfere with the synchronised coordination of cilia in the respiratory tract cannot really influence the flow of the cerebrospinal fluid. In our patient the total absence of kinocilia suggests that there may be a causal relation between primary ciliary dyskinesia and hydrocephalus.

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