Dialysis disequilibrium syndrome is the clinical phenomenon of acute neurological dysfunction attributed to cerebral edema that occurs during intermittent hemodialysis. We describe a case of DDS-induced intracranial hypertension in a patient with spina bifida and an adequately functioning shunt.

Case Report

This 19-year-old woman with a history of myelomeningocele closure and hydrocephalus since infancy was admitted to her local hospital for routine hemodialysis. Because of her age, she had recently been transferred to the local adult service from the regional pediatric nephrology unit to undergo dialysis treatment. During the initial dialysis treatment she developed severe headache, vomiting, and diplopia followed by tonic–clonic seizures. Given her history of hydrocephalus and a ventriculoatrial shunt in situ (peritoneal shunts had failed in the patient in the past due to adhesions and CSF malabsorption), she was urgently transferred to the regional neurosurgical unit for further evaluation.

On examination, the patient was of very small stature, weighing only 35 kg. She was afebrile, alert, and oriented with a Glasgow Coma Scale score of 15; she had no neurological deficits apart from long-standing paraplegia. Magnetic resonance imaging of the brain on admission revealed a well-decompressed ventricular system without evidence of cerebral edema despite her concurrent symptoms of raised ICP (Fig. 1 left). There were multiple MR imaging findings that were consistent with a Chiari malformation Type II including low-lying confluence of sinuses, a tubular elongated fourth ventricle, and a caudally displaced cervicomedullary junction. A CT scan of the brain obtained at the time of last shunt blockage (verified at surgery 1 year previously) revealed ventriculomegaly (Fig. 1 right).

Given her history of multiple shunt revisions, it was decided to monitor her ICP. The initial ICP readings were within normal range. During dialysis, however, the patient developed severe headaches, vomiting, diplopia, and ICP spikes of up to 50 mm Hg (Fig. 2). Following cessation of dialysis, the patient’s symptoms and intracranial hypertension gradually resolved over a 30-minute period. This pattern was replicated during subsequent dialysis treatment; the patient was completely asymptomatic and displayed normal-range ICP in the interdialysis interval.

Consultation with the regional nephrology service revealed that the patient received substantially different hemodialysis regimens in her local (adult) unit than in the children’s hospital (Table 1). Following adjustment of hemodialysis parameters, the patient’s symptoms during dialysis resolved and she has remained well on follow-up.

Abbreviations used in this paper: CSF = cerebrospinal fluid; DDS = dialysis disequilibrium syndrome; ICP = intracranial pressure.
Discussion

Dialysis disequilibrium syndrome was first reported in 1962 by Kennedy et al. It is characterized by a combination of neurological symptoms indicative of cerebral edema. Although the pathogenesis remains uncertain, the “reverse osmotic gradient” is thought to contribute. Urea removal occurs more slowly across the blood–brain barrier than from the plasma, generating an osmotic gradient that promotes water movement into the brain and cerebral edema. The rate of urea removal during dialysis may explain the association of DDS with hemodialysis and its absence in peritoneal dialysis when urea removal occurs more gradually over a longer time period. There are no reports in the literature to suggest that DDS is associated with peritoneal dialysis. One possible explanation is that peritoneal dialysis may allow more gradual equalization of urea and other osmo-

lytes across the blood–brain barrier, thereby minimizing any osmotic gradients.

Prevention is the mainstay of DDS treatment, particularly during initiation of hemodialysis in new patients. If unrecognized, cerebral edema refractory to treatment may develop, resulting in a fatal outcome. Prevention can be accomplished with intermittent hemodialysis using less efficient dialyzers with a smaller surface area and by reducing the duration and blood flow rate of dialysis.

Dialysis disequilibrium syndrome has been previously described in neurosurgical patients with head injury and intracerebral hemorrhage. To our knowledge ours is the first reported case in a patient with spina bifida and treated hydrocephalus. The diagnosis was confirmed by ICP monitoring during dialysis during which the patient experienced symptoms of intracranial hypertension, which correlated with high ICP readings. At the time of previous shunt failure, the patient’s images showed ventricular enlargement, which was absent on this occasion. Following adjustment of her hemodialysis parameters, her dialysis-related symptoms resolved. Failure of these measures to counteract the symptoms of DDS would be an indication for alternative modes of dialysis not associated with DDS (for example, peritoneal dialysis). However, peritoneal dialysis was contraindicated in our patient due to peritoneal malabsorption from adhesions related to multiple previous peritoneal shunts. Conversely, patients with peritoneal shunts should avoid peritoneal dialysis to minimize CSF malabsorption, and these patients may more appropriately undergo hemodialysis.

In our case, DDS may have been precipitated by the slit-like ventricles and lack of CSF buffer to compensate for cerebral edema development during dialysis. The radiological features of a Chiari malformation Type II would suggest an already “crowded” intracranial compartment, which may also have contributed to the development of DDS. Another explanation for the development of DDS relates to circulating blood volume in the venous system during hemodialysis. The absence of cerebral edema on MR images in our patient may lend support to this possibility. Many patients with spina bifida develop chronic renal impairment secondary to neuropathic bladder, and a proportion may eventually need hemodialysis. Such patients are often of small stature, making them vulnerable to large fluctuations in fluid and solute concentration during hemodialysis, particularly in the initial stages of treatment. Although we do not have weight measurements of our patient during dialysis, an increase in her circulating blood volume may have resulted in increased intracranial venous pressures. Furthermore, the low-lying confluence of sinuses present in many patients with spina bifida can be associated with high intracranial venous pressures, which result in an increase in resistance to CSF absorption and raised ICP. Careful titra-

FIG. 1. Left: Axial T2-weighted MR image of the brain showing the ventricular catheter in situ with well-decompressed ventricular system. Right: Brain CT scan (indicating ventriculomegaly) obtained 1 year previously at a time when the patient had a malfunctioning shunt.

FIG. 2. Tracings of ICP obtained before, during, and at the end of dialysis indicating an increase in ICP and return to normal pressure at the end of dialysis. The ICP recording was discontinued and recommenced while the patient was being transferred back to the ward from the dialysis unit. cns = central nervous system.

TABLE 1
Differences in the patient’s hemodialysis regimens between the adult and children’s dialysis centers

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Children’s Dialysis Unit</th>
<th>Adult Dialysis Unit</th>
</tr>
</thead>
<tbody>
<tr>
<td>duration of dialysis</td>
<td>2.25 hrs</td>
<td>2.5 hrs</td>
</tr>
<tr>
<td>blood flow during dialysis</td>
<td>300 ml/min</td>
<td>400 ml/min</td>
</tr>
<tr>
<td>dialyzer surface area</td>
<td>0.7 m²</td>
<td>2.1 m²</td>
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</tbody>
</table>
Dialysis disequilibrium syndrome in patients with hydrocephalus

The possibility of DDS should be considered in the evaluation of all shunt-treated patients who develop neurological symptoms during dialysis treatment.

Disclaimer
The authors do not report any conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

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