Sir,

With interest we read the paper of Wilcox et al. [1] on renal function in patients with cor pulmonale and secondary polycythaemia. Effective renal plasma flow (ERPF) was found to be reduced and filtration fraction (FF) to be increased. A few days after haematocrit (Hct) reduction, FF fell in 5 out of 7 patients. Parallel with the decrease in FF, a fall in body weight was seen. The authors suggest that, at least temporarily, the increased postglomerular capillary resistance – which is present in these patients [2] -can be corrected by Hct reduction.

We now show another effect of phlebotomy in a patient with congenital cyanotic heart disease. It is known that glomerular lesions can occur in these patients [3, 4]. Spear and Vitsky [3, 4] described glomerular enlargement, congestion and capillary dilatation. There was mesangial hyper-cellularity and focal glomerular sclerosis on light microscopy. On electron microscopy, diffuse thickening of the glomerular basement membrane was found [5]. Besides the abovementioned renal functional abnormalities also proteinuria is a common finding in these patients [4,5], probably as a consequence of the glomerular functional and morphological abnormalities.

We recently observed a 36-year-old man, known with tetralogy of Fallot, who underwent a Blalock operation at the age of 6. He was admitted to our hospital because of nephrotic syndrome and hypertension. Hct was 67%. Phlebotomy was carried out and he was instituted on digoxin. Body weight decreased and oedema disappeared but proteinuria persisted. From this period no accurate renal function studies are available. The patient was transmitted to the department of nephrology. At this moment no clinical signs of fluid overload were present and blood pressure was 140/95 mm Hg. Further investigations did not reveal any other abnormality known to cause nephrotic syndrome. Hct was 55%, proteinuria amounted to 4–5 g/

Table I. The effects of phlebotomy in our patient

<table>
<thead>
<tr>
<th>During After 8 weeks</th>
<th>observation phlebotomy 3, after discharge</th>
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</thead>
<tbody>
<tr>
<td>Body weight, kg</td>
<td>56.7  55.3  59</td>
</tr>
<tr>
<td>Haematocrit, %</td>
<td>55   44   59</td>
</tr>
<tr>
<td>Serum creatinine, µmol/l</td>
<td>86  77  91</td>
</tr>
<tr>
<td>Serum urea, mmol/l</td>
<td>7.7  5.0  5.3</td>
</tr>
<tr>
<td>GFR, ml/min</td>
<td>73  101  78</td>
</tr>
<tr>
<td>ERPF, ml/min</td>
<td>132  223  155</td>
</tr>
</tbody>
</table>
ERBF, ml/min 293 398 378
FF 0.55 0.45 0.50 Creatinine excretion 13,
mmol/24h 8.7 8.5 8.4
Albumin excretion 1*, g/24 h 4.5 2.2 4.7 Fractional albumin
excretion 13, Caib/CCreat
×102 0.15 0.06 0.13

a During 4 days 250 ml of blood was removed every day. Plasma was
reinfused. Measurements were performed the day after the last
phlebotomy.
b The mean of 3 days is given.

24 h, and serum albumin was 31 g/l (table I). Glomerular filtration rate (GFR) and ERPF [6]
were decreased with a striking increase in FF (normal value 0.25 ± 0.02). A kidney biopsy
showed glomerular enlargement, hypercellularity and sclerosis in the glomerular mesangium,
and hyalinization of afferent and efferent arterioles (fig. 1a). No tubular abnormalities were
found. Immunofluorescence showed weak linear deposition of IgA and IgG and slight granular
IgM and complement deposition. Electron microscopy revealed deposition of electron dense
material (possibly red cell fragments, globulins and clotting factors), along the endothelial aspect
of the glomerular capillary wall (fig. 1b).

The renal functional and morphological abnormalities in our patient are compatible with those
cited above [1–5]. It could be argued that the renal morphological abnormalities are secondary to
the polycythaemia, since high blood viscosity can induce increased postglomerular resistance
and thereby glomerular enlargement and capillary dilatation [2]. This could then initiate the
lesion of glomerular sclerosis [7]. Hct reduction in our patient was accompanied by a rise in GFR
and ERPF together with a fall in FF and proteinuria. This observation, therefore, underlines the
importance of a high transcapillary hydraulic pressure gradient in the development of glomerular
proteinuria (and sclerosis) in patients with congenital cyanotic heart disease.

Fig.1.a Expanded glomerulus with mesangial hypercellularity and sclerosis. Hilar arteriole shows
hyalinization. PAS stark. × 140. b Electron micrograph of glomerular capillary wall shows
accumulation of dense material (arrow heads) along the endothelial surface. × 11,000.

References
Wilcox, C.S.; Payne, J.; Harrison, B.D.W.: Renal function in patients with chronic hypoxaemia
Bauer, W.C.; Rosenberg, B.F.: A quantitative study of glomerular enlargement in children with
Spear, G.S.; Vitsky, B.H.: Hyalinization of afferent and efferent glomerular arterioles in cyanotic
Howenstine, J. A.; Lee, J. C.; Hopper, J.: The glomerular lesion of polycythaemia; in Vostal,