Visual outcome of malignant hypertension in young people

A C Browning, L S Mengher, R M Gregson, W M Amoaku

Abstract
A retrospective review was carried out of patients under 16 years old with malignant hypertension, who had been referred to a teaching hospital ophthalmology department because of reduced visual acuity. Four patients (three girls, one boy) were seen between 1994 and 2000 with a mean age at presentation of 11.5 years (range 9–15). In the short term, visual acuity improved after control of blood pressure in all four patients. However, in the long term, two patients were registered blind one to two years after presentation, one because of a choroidal neovascular membrane developing at the macula, and the other because of progressive optic neuropathy. Both of these patients had a longer duration of symptoms before diagnosis, worse visual acuity, and higher blood pressure at presentation when compared with the patients who made a good visual recovery. These observations suggest that early diagnosis of malignant hypertension in children is essential in reducing the likelihood of permanent severe visual damage.

(Keywords: malignant hypertension; vision; blind)

Malignant hypertension is defined as grade III hypertensive retinopathy in the presence of swollen optic discs (grade IV hypertensive retinopathy). Its incidence in children is unknown but it is generally accepted to be rare; while a range of predominantly reversible neurological and metabolic deficits have been documented, little is known about the long term visual prognosis associated with the optic nerve and chorioretinal changes. Of the small number of cases reported so far, the majority developed sudden blindness that was attributed to optic nerve ischaemia secondary to rapid blood pressure reduction.1–3 We present four children with malignant hypertension and document their clinical features and long term visual outcomes. Two were registered blind because of the delayed effects of the disease.

Case 1
A 9 year old girl was admitted to hospital after suffering a tonic–clonic seizure. This had been preceded by a 24 hour history of headache and vomiting. Six months earlier she had had a left Bell’s palsy, treated with prednisolone. She was also under investigation for nocturnal enuresis and vesico-ureteric reflux.

On examination, the patient had a blood pressure of 240/180 mm Hg (mean arterial pressure (MAP) 200 mm Hg); a diagnosis of hypertensive encephalopathy was made. Her visual acuity was recorded as 6/90 right, 6/240 left; fundoscopy revealed grossly swollen optic discs and macular stars. She was treated with intravenous labetolol and nitroprusside. Four weeks after presentation, her visual acuity was 6/24 right, 2/60 left. Her disc swelling was notably reduced but she still had extensive macular oedema and exudates. When seen one year after presentation, her visual acuity had reduced to 6/60 right, counting fingers (CF) left. The left macula showed extensive atrophic changes. After two years she developed a left relative afferent pupillary defect. The optic nerve disease was confirmed by visually evoked potentials.

She was registered blind three years after presentation, when her right visual acuity had reduced to 1/60 and the left eye was graded as perception of light (PL) only.

Case 2
A 12 year old girl was seen with a four week history of worsening vision and headaches. On examination, visual acuity was 6/60 right, 2/60 left. Dilated fundoscopy revealed bilateral optic disc swelling and macular stars. Her blood pressure was 220/180 mm Hg (MAP 193 mm Hg). She was admitted to hospital for control of her hypertension with oral nifedipine and propranolol. Investigation revealed her hypertension was secondary to reflux nephropathy. Six weeks after presentation, her visual acuity had improved to 6/24 bilaterally. Fundoscopy showed that her disc swelling had resolved but there were still notable macular exudates. After five months, her visual acuity remained unchanged but her macular exudates had resolved, leaving areas of hyper- and hypopigmentation.

Two years after presentation, she attended the eye department with a sudden decrease in visual acuity in the left eye to CF. Fundoscopy showed an area of macular subretinal elevation with associated haemorrhage. Clinically this was thought to represent an area of subfoveal choroidal neovascularisation, subsequently confirmed by fluorescein angiography (fig 1). At this stage her blood pressure was well controlled with enalapril and her renal function was stable with a plasma creatinine of 99 µmol/l. Six months later, the lesion had evolved into a disciform scar (fig 2). One year later she was registered blind after her right eye deteriorated to a visual acuity of 1/60 because of atrophic maculopathy. Her optic discs remain healthy and her visual acuity remains stable at 1/60 right, CF left, four years after presentation.
Case 3
A 10 year old boy presented with a three week history of blurred vision and headache. He was noted to have bilateral macular stars and swollen optic discs (fig 3). His visual acuity at presentation was 6/24 right, 6/36 left and his blood pressure was recorded as 178/126 mm Hg (MAP 143 mm Hg). A diagnosis of malignant hypertension was made and he was treated with sublingual nifedipine and intravenous labetolol. Subsequent investigation revealed acute on chronic renal failure secondary to probable reflux nephropathy. One month after presentation his visual acuity had improved to 6/12 right, 6/9 left, although some disc swelling and macular exudate were still present in both eyes. By seven months his visual acuity had improved to 6/9 right, 6/6 left and his optic disc swelling and macular exudates had resolved to reveal dark pigmentary stippling scattered over each macula.

Case 4
A 15 year old girl was admitted to hospital after suffering three tonic–clonic seizures over a period of four hours. These were preceded by a 24 hour history of increasing disorientation and headache. On admission, her blood pressure was 240/135 mm Hg (MAP 170 mm Hg) and the diagnosis of hypertensive encephalopathy was made. She was ventilated and treated with intravenous labetolol. Fundoscopy revealed bilateral swollen optic discs and macular exudates.

Six days after admission she was seen in the ophthalmology clinic where she gave a history of four weeks of intermittent blurred vision and headaches. Her visual acuity was 6/9 right, 6/5 left. Fundoscopy showed residual bilateral macular exudates but normal optic discs. After six months her visual acuity had improved to 6/6 right and 6/6 left. The left macula appeared healthy but that on the right still had residual exudates. By nine months, her visual acuity was 6/5 in both eyes. Fundoscopy of the right eye showed a small area of macular retinal pigment epithelium hyperpigmentation. Investigation had shown her to have chronic renal failure secondary to vesicoureteric reflux.

Discussion
Hypertension in children has an incidence of approximately 1–2%. However, prevalence of malignant hypertension is unknown but generally accepted as being extremely rare. It may be associated with non-specific features such as reduced conscious level, seizures, headache, persistent vomiting, transient hemipareses, and VII nerve palsy.7 The patient may also present to ophthalmologists with symptoms that include reduced visual acuity secondary to either hypertensive retinopathy, choroidopathy, or optic neuropathy, or because of cortical disease causing transient loss of colour vision, visual hallucinations, and ocular motility disorders.7

Most reports of visual impairment associated with malignant hypertension have reported precipitous loss of vision associated with too rapid a reduction of blood pressure, leading to optic nerve infarction.8 A number of authors have stated the dilemma of the need to reduce blood pressure quickly to avoid permanent central nervous system, cardiac, and renal damage but at the same time not exposing watershed areas of the brain and optic nerves to irreversible ischaemic damage.6

Importantly, the blood pressure of our patients was reduced in line with current guidelines for the treatment of malignant hypertension in children.8 These suggest that a reduction in blood pressure by one third of the total reduction planned is reasonable during the first six hours, a further third over the next 12–36

Figure 1  Fluorescein angiogram of the left eye of case 2, showing subfoveal leakage of dye corresponding to the choroidal neovascular membrane.

Figure 2  Fundal photograph of the left eye of case 2, showing a pigmented macula disciform scar.

Figure 3  Fundal photograph of the left eye of case 3, showing a swollen disc, multiple cotton wool spots, arterial attenuation, and a macula star.
hours, and the final third slowly over the following 48–96 hours.

Logan et al reported the visual outcome of three children diagnosed with malignant hypertension. All had a permanent reduction in visual acuity ranging between 6/9 and 6/36. All three cases had healthy optic discs but showed atrophy at the macula, ascribed to chronic macular oedema.

Our four cases show the wide range of potential ophthalmic outcomes in children with malignant hypertension. In two (cases 3 and 4), all signs resolved except some pigmentary changes at the maculae and visual acuity returned to normal in both. Their symptoms were of short duration and they had good initial visual acuity (6/5 to 6/36). Their mean arterial blood pressures at presentation were 173 and 143 mm Hg respectively.

Two children suffered a delayed fall in visual acuity, eventually resulting in blindness. Case 1 presented with a BP of 240/180 (MAP 200 mm Hg) and had premorbid symptoms of up to six months. She had very poor vision at presentation and suffered a gradual decline to 1/60 right and PL left over two years, despite optimal blood pressure control. Case 2 presented with a four week history of symptoms and a maximum recorded BP of 220/180 (MAP 193 mm Hg). She had an initial improvement in visual acuity from 6/60 right, 2/60 left at presentation to 6/24 both eyes by six weeks. However, two years after presentation her vision in one eye decreased suddenly from a choroidal neovascular membrane and later this occurred in the other eye. In the absence of inflammation, the most likely cause underlying the neovascular membrane was chronic ischaemia of the choriocapillaris.

Although small, this case series illustrates a number of points. Generally the worst visual prognosis was associated with the highest presenting blood pressure, the worst visual acuity at presentation, and the longest duration of symptoms. Two cases suffered a significant drop in visual acuity between one and two years from presentation despite satisfactory initial and subsequent blood pressure control, one from progressive optic neuropathy and the other from a choroidal neovascular membrane. To our knowledge, this is the first case of choroidal neovascularisation reported in a child with malignant hypertension.

These cases illustrate that when treating children with malignant hypertension we should be aware that significant deterioration in vision may occur many months after the initial apparent success of treatment. It is likely that visual prognosis is affected by factors present at the time of presentation, so that early recognition is essential. This can only be achieved by having a low threshold for blood pressure measurement in children presenting with atypical neurological or visual symptoms, particularly if there is a history of vesicoureteric reflux.

Perhaps the most important lesson of this case series is the need to prevent the occurrence of reflux nephropathy. This requires early identification and thorough treatment of infants with urinary tract infections, and subsequent long term follow up.

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