Peripheral facial nerve palsy typically presents with weakness of all muscles of the face: The facial creases and nasolabial fold disappear, the forehead unfurrows, and the corner of the mouth droops. In more severe cases, the eyelid does not close and on attempted closure the Bell’s phenomenon with the eye rolling upward becomes noticeable. Acute peripheral facial nerve palsy may result from infectious conditions such as Lyme disease, otitis media, chickenpox, herpes zoster oticus, mumps, and mononucleosis; from noninfectious, presumably inflammatory disorders such as the Melkersson-Rosenthal syndrome; and from traumas or neoplasms. However most cases present without an apparent cause and are classified as Bell’s palsy.1 There is also an association between peripheral facial nerve palsy and severe systemic hypertension, as was first described by Moxon more than a century ago.2-3 We systematically review the literature on this entity and report on two children in whom facial nerve palsy led to the detection of severe systemic hypertension.

METHODS
The US National Library of Medicine, Embase, Web of Science, and the web-based search engine Google Scholar were searched using the terms arterial hypertension, systemic hypertension, and malignant hypertension, each combined with peripheral facial nerve paralysis and Bell paralysis, limiting the search to publications in English, German, and Italian dating from January 1960 to December 2011. If the publication was available as a full-length article or a letter, if the patients included were individually described revealing at least age, sex, and blood pressure (BP) measurement in the hypertensive range; if the facial palsy was specified as being of the peripheral type, the publication was included in this study. From each case of severe systemic hypertension complicated by peripheral facial nerve palsy, two authors (R.J. and G.P.M.) independently excerpted the patients’ clinical characteristics including information on the underlying cause of systemic hypertension, family and previous

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personal history, further investigations performed, such as neuroimaging, treatment modalities applied, and critically appraised the publications.

The time to diagnosis was defined as the time elapsed between the first facial symptoms and the diagnosis of arterial hypertension. Severe hypertension was defined as a BP exceeding the 99th percentile by >5 mm Hg in children or 180/110 mm Hg in adults. For each patient the extent of severe hypertension was expressed by means of the modified BP severity index, which is calculated dividing the measured BP (in mm Hg) by either the calculated 99th percentile BP value specific for that patient plus 5 mm Hg in children or by 180/110 mm Hg in adults. The facial nerve dysfunction was graded as mild in subjects with preserved eye closure and severe in those with impaired eye closure.

Written informed consent for publication of information regarding the two described cases was obtained from both patients’ parents.

Continuous data are presented as median values with interquartile range (IQR) or minimum and maximum range, and categorical data as frequencies and percentages calculated from the results reported in the original publications. Correlation analyses were performed with the Spearman test. Comparison of prevalence between the groups was performed using the Fisher exact test. All the statistical analyses were performed with GraphPad Prism, version 4.01 for Windows (GraphPad Software, San Diego, CA).

RESULTS

Systematic review of the literature

The systematic review revealed 326 publications, of which 282 remained after excluding duplicates. Fifteen publications met the inclusion criteria. The references of these 15 reports revealed three further publications meeting the inclusion criteria. In total, 18 reports were included in this study (17 in English, one in Italian) describing 24 cases. All publications included were reports of single cases or case series.

Finally, we found 41 additional cases of peripheral facial nerve palsy in five publications which specifically addressed the neurological signs of severe systemic hypertension in pediatric patients. These cases were only listed and not described in detail, permitting only analysis of prevalence.

Case reports

Case 1. A previously healthy 5-year-old boy developed left facial muscular weakness, which spontaneously recovered over two months. At this time BP was not measured. Three months later he was referred because of headache, nocturia, and a similar facial weakness on the same side. The physical examination disclosed left-sided peripheral facial nerve paresis with normal facial symmetry and tone at rest, four-limb BP of 220/160 mm Hg, and bilateral retinal hemorrhages and tortuous vessels on fundoscopy. Urinalysis disclosed pathological proteinuria (+/++) and mild erythrocyturia. Electrocardiography and echocardiography revealed left ventricular hypertrophy. Blood investigations showed a normal complete blood cell count and renal profile, a normal circulating complement C3 level, and absent antinuclear and antineutrophil autoantibodies. The final diagnosis of segmental renal hypoplasia with systemic hypertension was made by means of renal ultrasound and, subsequently, contrast-enhanced abdominal computed tomography, which revealed a segmental cortical thinning and dilated calices in the upper part of the right kidney. A voiding cystourethrography failed to detect a vesicoureteral reflux. The BP was gradually brought under control, initially with intravenous labetolol, later on medication with ramipril and chloralidon. After 2 weeks, BP values were slightly below the 95th percentile (108/69 mm Hg). The facial nerve paresis resolved after BP was brought under control and completely within 2 months. One year later, BP remained within normal range and further neurological complications were not observed.

Case 2. In a previously healthy 11-year-old girl with sudden onset of left facial weakness and negative serologic testing for Lyme disease, the suspected diagnosis of idiopathic peripheral facial nerve paresis was made and oral prednisone (1 mg per kg body weight per day for 7 days) prescribed. At this time BP was not measured. Ten days later the girl experienced a generalized tonic-clonic seizure and was admitted to hospital. Physical examination revealed a left-sided peripheral facial nerve paresis (with normal facial symmetry and tone at rest), four-limb BP of 230/170 mm Hg, bilateral retinal hemorrhages and tortuous vessels on fundoscopy, and an epigastric systolic bruit. Urinalysis disclosed pathological proteinuria (+++/+++) and erythrocyturia. Electrocardiography and echocardiography revealed a left ventricular hypertrophy. Blood investigations showed a normal complete blood cell count and renal profile, and a negative autoimmune profile. The BP was gradually brought under better control (after 1 week BP was 123/84 mm Hg), initially with intravenous labetolol, later on combined medication with amlodipine and chloralidon. Renal angiography disclosed a proximal left renal artery stenosis secondary to a fibromuscular dysplasia that was successfully treated by angioplasty. The facial nerve paresis resolved within 6 weeks, after having brought BP under control (at that time BP was 115/74 mm Hg). On follow-up, after 6 months, antihypertensive treatment could be tapered off and BP remained within the normal range 1 year later.

Analysis of all cases

Demographics. Of the 24 patients reported in the literature and the presented two cases with severe systemic hypertension complicated by peripheral facial nerve palsy, 16 were female, 10 male. Median age was 9.5 years (IQR, 2.5–12 years; range, 11 weeks–62 years). Twenty-three patients (88%) were children 15 years of age or younger (Table 1, Figure 1). The mean modified severity index of arterial BP was 1.67 (IQR, 1.39–1.73)/1.58 (IQR, 1.36–1.84).

Characteristics of peripheral facial palsy. At presentation the facial palsy was unilateral in 25 cases (left-sided, n = 14;
Facial Nerve Palsy in Hypertension

Table 1. Characteristics of 26 patients with severe hypertension and peripheral facial nerve paresis

<table>
<thead>
<tr>
<th>Reference number</th>
<th>Age (years)</th>
<th>Gender</th>
<th>Additional presenting symptoms</th>
<th>Blood pressure (mm Hg)</th>
<th>Underlying disease</th>
<th>Treatment other than antihypertensive medication</th>
<th>Neuroimaging</th>
<th>Recurrence of facial palsy</th>
<th>Time to resolution of palsy</th>
<th>Resolution of palsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case I</td>
<td>5</td>
<td>M</td>
<td>H</td>
<td>220/160</td>
<td>CAKUT</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>2 months</td>
<td>Complete</td>
</tr>
<tr>
<td>Case II</td>
<td>11</td>
<td>F</td>
<td>C</td>
<td>230/170</td>
<td>Renal artery stenosis</td>
<td>Renal angioplasty</td>
<td></td>
<td></td>
<td>1.5 months</td>
<td>Complete</td>
</tr>
<tr>
<td>5</td>
<td>47</td>
<td>M</td>
<td>A, O, V</td>
<td>216/142</td>
<td>Primary hypertension</td>
<td>No</td>
<td>MRI: Ischemic infarct of dorsal pons</td>
<td></td>
<td>?</td>
<td>?</td>
</tr>
<tr>
<td>6</td>
<td>9</td>
<td>F</td>
<td>A, C, H, O, V</td>
<td>200/150</td>
<td>CAKUT</td>
<td>No</td>
<td>MRI: Pontine hemorrhage</td>
<td>Yes</td>
<td>0.5 months</td>
<td>Complete</td>
</tr>
<tr>
<td>7</td>
<td>5</td>
<td>M</td>
<td>A</td>
<td>260/175</td>
<td>Pheochromocytoma</td>
<td>No</td>
<td>CT: No acute changes, persistent chronic lesions</td>
<td>Yes</td>
<td>Died after 4 months</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>62</td>
<td>F</td>
<td>—</td>
<td>220/98</td>
<td>Renal artery stenosis</td>
<td>No</td>
<td>CT: No acute changes, persistent chronic lesions</td>
<td>Yes</td>
<td>0.5 months</td>
<td>Almost complete</td>
</tr>
<tr>
<td>9</td>
<td>11</td>
<td>F</td>
<td>H, F, V</td>
<td>215/135</td>
<td>Primary hypertension</td>
<td>No</td>
<td>MRI: Hypertensive encephalopathy</td>
<td>Yes</td>
<td>0.5 months</td>
<td>Complete</td>
</tr>
<tr>
<td>10</td>
<td>14</td>
<td>F</td>
<td>H</td>
<td>230/150</td>
<td>Renal adenocarcinoma</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>1 month</td>
<td>Complete</td>
</tr>
<tr>
<td>11</td>
<td>10</td>
<td>M</td>
<td>H</td>
<td>140/100</td>
<td>Guillain-Barré syndrome</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>6 months</td>
<td>Complete</td>
</tr>
<tr>
<td>12</td>
<td>42</td>
<td>F</td>
<td>—</td>
<td>200/150</td>
<td>Primary hypertension</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>?</td>
<td>?</td>
</tr>
<tr>
<td>13</td>
<td>0.9</td>
<td>F</td>
<td>—</td>
<td>200/160</td>
<td>Mid aortic syndrome</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>6 months</td>
<td>Complete</td>
</tr>
<tr>
<td>14</td>
<td>1</td>
<td>M</td>
<td>—</td>
<td>240/130</td>
<td>Malignant hypertension of unknown origin</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>2 months</td>
<td>Complete</td>
</tr>
<tr>
<td>15 – I</td>
<td>0.2</td>
<td>M</td>
<td>—</td>
<td>133/94</td>
<td>Coarctation</td>
<td>Surgical correction</td>
<td>MRI: Normal</td>
<td>Few days</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>15 – II</td>
<td>13</td>
<td>M</td>
<td>—</td>
<td>218/122</td>
<td>Coarctation</td>
<td>Surgical correction</td>
<td>MRI: Normal</td>
<td>Few days</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>0.3</td>
<td>M</td>
<td>—</td>
<td>200/-</td>
<td>Coarctation</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>3 weeks</td>
<td>Complete</td>
</tr>
<tr>
<td>17 – I</td>
<td>10</td>
<td>F</td>
<td>—</td>
<td>190/130</td>
<td>Primary hypertension</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>Several months</td>
<td>Complete</td>
</tr>
<tr>
<td>17 – II</td>
<td>7</td>
<td>F</td>
<td>—</td>
<td>192/120</td>
<td>CAKUT</td>
<td>Nephrectomy</td>
<td></td>
<td>3 months</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>17 – III</td>
<td>10</td>
<td>F</td>
<td>O</td>
<td>205/153</td>
<td>CAKUT</td>
<td>No</td>
<td>MRI: Multiple ischemic bilateral insular, occipital and basal ganglia infarcts</td>
<td>9 months</td>
<td>Almost complete</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>4</td>
<td>M</td>
<td>O</td>
<td>270/160</td>
<td>Renal artery stenosis</td>
<td>Surgical correction</td>
<td>MRI: Ischemic nucleus lentiformis and periven- tricular infarcts</td>
<td>Yes</td>
<td>12 months</td>
<td>Complete</td>
</tr>
<tr>
<td>19 – I</td>
<td>5</td>
<td>F</td>
<td>H, O</td>
<td>130/100</td>
<td>Guillain-Barré syndrome</td>
<td>IVIG</td>
<td>MRI: Enhancement of the lumbosacral spinal nerve roots</td>
<td>4 months</td>
<td>Partial</td>
<td></td>
</tr>
<tr>
<td>19 – II</td>
<td>3</td>
<td>F</td>
<td>O</td>
<td>126/110</td>
<td>Guillain-Barré syndrome</td>
<td>IVIG</td>
<td>MRI: Enhancement of the lumbosacral spinal nerve roots</td>
<td>3 months</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>20 – I</td>
<td>12</td>
<td>F</td>
<td>V</td>
<td>220/140</td>
<td>CAKUT</td>
<td>No</td>
<td>CT: Normal</td>
<td>6 months</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>20 – II</td>
<td>13</td>
<td>F</td>
<td>—</td>
<td>200/140</td>
<td>CAKUT</td>
<td>No</td>
<td>MRI: Normal</td>
<td>8 months</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>21 – I</td>
<td>0.7</td>
<td>F</td>
<td>C</td>
<td>220/170</td>
<td>CAKUT</td>
<td>Nephrectomy and facial nerve decompression</td>
<td></td>
<td>1 months</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>21 – II</td>
<td>12</td>
<td>F</td>
<td>H, V</td>
<td>230/170</td>
<td>?</td>
<td>No</td>
<td></td>
<td>Several weeks</td>
<td>Complete</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>1</td>
<td>M</td>
<td>C</td>
<td>220/120</td>
<td>?</td>
<td>No</td>
<td></td>
<td>Yes</td>
<td>1 months</td>
<td>Complete</td>
</tr>
</tbody>
</table>

Abbreviations: A, altered mental status; C, convulsions; CAKUT, congenital anomalies of the kidney and urinary tract; CT, computed tomography; F, fatigue; H, headache; IVIG, intravenous immunoglobulines; MRI, magnetic resonance imaging; O, other central neurological manifestations; V, vomiting.

*Sixteen female and 10 male subjects, aged between 0.2 and 62 years (median, 9.5 years).
In the three children with Guillain-Barré syndrome, both aortic coarctation, renal artery stenosis, consisting of congenital anomalies of the kidney and urinary tract, and severe hypertension were the presenting signs before further features of the polyneuropathy were apparent. The cause of arterial hypertension remained unknown or classified as primary hypertension in seven patients.

Concomitant signs and symptoms. Additional signs of nervous system dysfunction were described in 16 patients, consisting of headache (n = 7), vomiting (n = 5), convulsions (n = 4), hyperactive reflexes (n = 2), positive Babinski sign (n = 1), altered level of consciousness (n = 3), visual field defect (n = 1), internuclear ophthalmoplegia (n = 1), and not further described transient double vision (n = 1), as depicted in Table 1. In three patients, peripheral facial nerve palsy and severe systemic hypertension were associated with Guillain-Barré syndrome. In one patient, computed tomography of the brain revealed a pontine hemorrhage at the site of the facial nucleus in a 9-year-old girl, and an ischemic stroke in the dorsal pons affecting the post-nuclear facial fibers in a 47-year-old man. Four patients had signs of multiple ischemic infarcts in the periventricular white matter; three showed involvement of the thalamus and the basal ganglia, one had additional lesions in the insular and occipital cortex. In two patients with Guillain-Barré syndrome and gadolinium enhancement of spinal nerve roots on MRI, no cerebral or cranial nerve abnormalities were described. In three patients, one of whom had a computed tomography, neuroimaging was normal (Table 1).

No electrophysiological findings specifically addressing the facial nerve were reported.

An edematous nerve sheath with vessel engorgement without hemorrhage was revealed in the child who underwent decompression of the facial nerve from the cochleariform process to the stylomastoid foramen.

Therapeutic approaches and outcome. In 23 patients, BP reduction after correction of the underlying disorder or antihypertensive drug management was followed by a gradual resolution of the facial nerve palsy (19 complete and four partial resolutions, median time to resolution was 2 [IQR, 0.9–6] months; Table 1). In an 8-month-old girl facial nerve palsy resolved following surgical removal of a nonfunctioning kidney, medical stabilization of BP and surgical decompression of the facial nerve. The child with persistent facial nerve palsy reported by Bialestock in 1961 died because of very severe, refractory hypertension.

Steroids were prescribed in three cases reported in the literature and in our case II. All four of these patients were children in whom BP had not been measured at initial diagnosis of peripheral facial nerve palsy, which had been interpreted as Bell’s palsy and subsequently treated with corticosteroids. No information on dosage and duration of steroid treatment was available in the three patients found in the literature.

Peripheral facial nerve palsy in publications dealing with nervous system involvement in the context of severe hypertension

Forty-one cases of peripheral facial nerve palsy were found in five publications which specifically addressed the neurological signs of systemic hypertension in a total of 860 severely hypertensive children (4.8%, Table 2). Headache was by far the most common neurological complication in children with severe arterial hypertension, followed in decreasing order by altered level of consciousness and vomiting, convulsions, focal central nervous system deficit, and peripheral facial nerve palsy.
Table 2. Peripheral facial nerve palsy noted in five pediatric case series with a total of 860 patients affected by severe hypertension

<table>
<thead>
<tr>
<th>First Author</th>
<th>Leumann23</th>
<th>Trompeter25</th>
<th>Deal26</th>
<th>Rance14</th>
<th>Lloyd14</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of patients</td>
<td>600</td>
<td>45</td>
<td>110</td>
<td>70</td>
<td>35</td>
<td>860</td>
</tr>
<tr>
<td>Patients with facial nerve paralysis (%)</td>
<td>20 (3.3)</td>
<td>2 (4.4)</td>
<td>10 (9.1)</td>
<td>2 (2.8)</td>
<td>7 (20)</td>
<td>41 (4.8)</td>
</tr>
</tbody>
</table>

These cases were only listed and not described in detail, permitting only analysis of prevalence.

DISCUSSION

The main finding of this study is the fact that peripheral facial nerve palsy associated with severe arterial hypertension is primarily described in children and adolescents. The long time to diagnosis of arterial hypertension, lasting a median of 45 days, indicates that this association is not widely recognized. Particularly in children, BP measurement is not firmly considered as a critical component of the routine physical examination. However, recent clinical reviews addressing the management of peripheral facial palsy in childhood mention severe systemic hypertension as a specific cause.1, 27 Within the group of severely hypertensive children and adolescents, peripheral facial palsy was described in 4.8% (41/860) ranging from 2.8% to 20% as indicated in five publications dealing with the complications of severe childhood hypertension.4, 23–26 Moreover, in a survey dealing with the different etiologies of peripheral facial palsy, arterial hypertension was the cause in 7 of 87 (8%) children.14

Lower motor neuron facial palsy has previously been associated with the hypertensive disorders of pregnancy and puerperium,28 and a recent case-control study suggests that chronic, nonsevere hypertension may increase the risk of lower motor neuron facial nerve palsy in subjects older than 40 years of age.29

A timely diagnosis of arterial hypertension in the case of peripheral facial palsy is crucial in order to start adequate antihypertensive treatment. The majority of the cases showed full recovery of the facial palsy some weeks after onset of treatment. It remains unclear whether medication directly affects speed and degree of resolution or acts only to prevent further recurrence, since some cases demonstrated spontaneous resolution prior to the diagnosis of arterial hypertension. In the three adult cases with known chronic arterial hypertension, facial palsy occurred during exacerbation of severe arterial hypertension due to nonadherence of medication and immediately resolved after better BP control, suggesting a direct relationship between BP lowering and resolution of facial palsy.5,8,12 Corticosteroids, the standard therapy for Bell’s palsy, potentially exacerbate arterial hypertension and are therefore not indicated.

The putative mechanisms underlying the dysfunction of the facial nerve or its nucleus were addressed in a minority of the affected patients. These include a swelling of the facial nerve in its bony canal associated with vessel engorgement, a hemorrhage into the facial canal, as reported in two necropsies,3, 30 an ischemic stroke affecting the postnuclear fibers of the nerve, or a bleeding in the facial nerve nucleus. These mechanisms may be implicated also in the development of facial palsy during preeclampsia.28

Both lower motor neuron facial nerve palsy and arterial hypertension have independently been described as features of Guillain-Barré syndrome.31 Whether the cases of facial nerve palsy reported in three children with Guillain-Barré syndrome included in this analysis were caused by severe hypertension or were a first manifestation of the polyneuropathy remains uncertain.

The most important limitation of this review lies in the small number of reported patients affected by peripheral facial nerve palsy caused by severe systemic hypertension and the retrospective nature of the analysis. A second limitation is that adequate neuroimaging studies were rarely performed. Finally, the degree of facial palsy was not assessed using a facial nerve grading system.

In conclusion, this review provides evidence that there might be an association of uncontrolled hypertension and facial palsy that warrants additional evaluation. With effective acute and chronic antihypertensive therapy, the prognosis of facial palsy is excellent. A gradual normalization of BP in 2–3 days is advised. 5,32–33

DISCLOSURE

The authors declared no conflict of interest.

REFERENCES