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EXTENSIVE CLINICAL EXPERIENCE

Primary Empty Sella

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Context: The term primary empty sella (PES) refers to a number of endocrine and/or neurological disturbances that may be caused by the herniation of subarachnoid space within the sella.

Setting: The records of all patients with a diagnosis of empty sella between 1985 and 2002 seen at the Catholic University of Rome and University of Brescia were examined retrospectively.

Patients: We have observed 171 female and 42 male patients affected by PES (over 4:1 sex ratio). The mean age at diagnosis in our subjects was 51.8 ± 2.1 yr. Mean body mass index was 27.3 ± 3.5 kg/m².

Main Outcome Measure: All the patients have been analyzed first either with sellar computed tomography scan or magnetic resonance imaging. All patients underwent neurological, ophthalmological, and baseline endocrine evaluation (appropriate stimulation tests were performed when hypopituitarism was suspected).

Results: In the overall population, 40 of 213 patients had documented endocrine abnormalities, specifically 31 females and nine males. Twenty-two patients (10.3% of total patients; 18 women, 10.5% of all women, with a mean age of 38.6 ± 1.1 yr and four males, with a mean age 46.5 ± 3.52 yr) presented with hyperprolactinemia. Global anterior hypopituitarism was confirmed in nine patients. Eight patients presented an isolated GH deficiency. One hundred thirty-eight of our patients presented a so-called partial empty sella at computed tomography scan/magnetic resonance imaging, and 75 had total PES.

Conclusions: PES may be associated with variable clinical conditions ranging from mild endocrine disturbances to severe intracranial hypertension and rhinorrhea. The need for treatment of hyperprolactinemia as well as for replacement hormone therapy must be assessed in PES. Symptomatic intracranial hypertension makes cerebrospinal fluid shunting procedures necessary. (J Clin Endocrinol Metab 90: 5471–5477, 2005)

EMPTYSSELLA IS characterized by the herniation of the subarachnoid space within the sella, which is often associated with some degree of flattening of the pituitary gland (1, 2).

In the case of primary empty sella (PES), several etiopathogenetic hypotheses have been proposed, including a congenital incomplete formation of the sellar diaphragm and supra sellar factors such as stable or intermittent increase in intracranial pressure as well as volumetric changes in the pituitary (as observed in pregnancy) (1–3). On the other hand, secondary empty sella may be caused either by pituitary adenomas undergoing spontaneous necrosis (ischemia or hemorrhage) or by infective, autoimmune, and traumatic causes or by radiotherapy, drugs, and surgery.

Several series of patients with PES have been reported in the last two decades in the literature (3–7). However, to our knowledge, no large enough series have been reported so far based on a single or a few centers’ experience to allow a detailed definition of the epidemiological, diagnostic, and therapeutic picture of patients with PES.

The aim of our paper was to report the clinical experience of two Italian endocrine centers, which are referral centers for pituitary diseases of large areas in Northern (Brescia) and Central (Rome) Italy. Because of the significant number of patients evaluated (n = 213), insights on clinical aspects and outcome of PES may be gained. Based on our report, we will also attempt to draw some conclusions that may point toward a diagnostic and therapeutic approach as well as the follow-up of patients with PES.

Patients and Methods

Selection of patients

The records of all patients with a diagnosis of empty sella between 1985 and 2002 seen at the Department of Endocrinology and Neurosurgery of the Catholic University of Rome and at the Endocrine section, Department of Internal Medicine, University of Brescia, were examined retrospectively, 1 yr or more after hospital discharge, to ensure a reasonable follow-up time. These clinical records were summarized into an ad hoc designed questionnaire in which anthropometric and historical data together with all endocrine, neurological, ophthalmological, and radiological available data of each patient at diagnosis and during follow-up were included. Criteria for exclusion from the data collection were 1) known history of either hypothalamic-pituitary or central nervous system diseases; 2) previous medical, neurosurgical, and radiation treatment for pituitary adenomas; and 3) ascertained GH and cortisol hypersecretion or patients with prolactin levels greater than 100 ng/ml in whom the possibility of an empty sella secondary to the necrosis of a preexisting pituitary adenoma could not be ruled out. Therefore, the present study was based on the analysis of the records of 213 patients who were likely bearing a PES, although it has to be noted that this diagnosis remains in most cases presumptive because the above mentioned criteria allow us to consider highly unlikely but not to completely rule out the possibility that in a few patients asymptomatic infarction of a previously undiagnosed pituitary tumor may have occurred.
Methods

All the patients were analyzed first with either sellar computed tomography (CT) scan or magnetic resonance imaging (MRI). All the patients underwent neurological and ophthalmological evaluation and baseline endocrine evaluation (fasting morning serum samples for free \( T_3 \) and \( T_4 \), TSH, ACTH, cortisol, 24-h urinary cortisol, IGF-I, LH, FSH, 17β-estradiol, and testosterone). Samples for prolactin assay were obtained after 40 min of iv saline infusion with the patient recumbent. Appropriate stimulation tests [TRH (Serono, Rome, Italy), 200 \( \mu \)g iv bolus injection with blood samples for TSH assay taken at −15, 0, and 20, 40, and 60 min after stimulation (8); GHRH (Geref; Serono), 1 \( \mu \)g/kg iv bolus injection with blood samples for GH assay taken at −15, 0, and 15, 30, 45, 60, 90, and 120 min after stimulation (9, 10); and GnRH (Lutrelef; Ferring, Germany), 100 \( \mu \)g iv bolus injection with serum samples for the determination of LH and FSH levels taken at −15, 0, and 15, 30, 45, 60, 90, and 120 min after the stimulation (11)] were performed when hypopituitarism was suspected (n = 36 subjects). Standard immunoradiometric assays and RIAs have been used for hormone level detection. Results are always expressed as mean ± SEM.

Results

Epidemiology and risk factors

We have observed 171 female and 42 male patients affected by PES (over 4:1 sex ratio). The mean age at diagnosis in our subjects was 51.8 ± 2.1 yr. The peak incidence occurred between the ages of 30 and 41, being somewhat earlier in women than in men, perhaps because of the greater frequency of pituitary examination in women with oligomenorrhea (Fig. 1). Autoimmune hypothyroidism was present in five patients. Of the female patients with PES, only 19 did not undergo at least one pregnancy. The remaining population mean number of pregnancies was 1.96 ± 0.1. In detail, about 57% of these women were multiparous (53, 23, and nine patients had, respectively, two, three, and four pregnancies). Most of the patients (155 of 213, 73%) were overweight [body mass index (BMI) > 25 kg/m\(^2\)]. Some of the patients were significantly obese (BMI > 30 kg/m\(^2\)) (25 females, i.e. 15% of total female patients, with a mean age of 37.6 ± 2.84 yr; five males, i.e. 12% of total male patients, with a mean age of 30.0 ± 6.27 yr). The majority of these patients (18 of 30) had respiratory problems such as sleep apnea syndrome and Pickwick syndrome. Mean BMI of the entire population was 27.3 ± 3.5 kg/m\(^2\). In Fig. 2, distribution of the studied population according to BMI is reported. In our patients, neither type 1 diabetes mellitus nor primary hypoadrenalism nor hypoparathyroidism were observed.

Empty sella is reported in a range between 5.5% (12) and 23% (13) in the autopsic examination of the sellar region, and this condition is not uncommon, being reported in the 8–35% of the general population (14–17). In the past, it has been shown that PES is more frequent in female subjects (5, 18). Our data confirm a significant prevalence of PES in females.

PES has also been reported in association with several endocrine autoimmune diseases, and PES itself has been suggested to be a consequence of lymphocytic hypophysitis (19). Our data suggest that autoimmunity per se may not be significantly linked to the occurrence of PES (20).

On the other hand, pregnancy could promote the onset of PES (21, 22). In fact, the pituitary volume could double during pregnancy, particularly in the case of multiple pregnancies (23). This may contribute to cause the herniation of subarachnoid space in case of hypoplastic diaphragma sellae and/or cerebrospinal fluid (CSF) hypertension, even if moderate and temporary. Our data show a large prevalence of PES among women with at least one completed pregnancy in their physiological history. However, to demonstrate the role of pregnancy in the pathogenesis of PES, comparison with a control group would be needed.

PES has also been reported to be associated with obesity. In fact, it is believed that morbid obesity may induce hypercapnia which can be the cause of chronic CSF pressure elevation and in turn may lead, in subjects with hypoplastic diaphragma sellae, to the intrasellar herniation of the suprasellar subarachnoid space (3, 18). Our data suggest that morbid obesity may play a marginal role, if any, in the pathogenesis of empty sella because the prevalence of this phenotype does not appear to be clearly elevated in our PES population with respect to the general population.

Finally, other risk factors such as type 2 diabetes, hypertension (21), use of certain drugs (5, 24), and history of pseudotumor cerebri have been proposed in patients with PES. We were not able to find an increased prevalence of these factors in our patients with respect to the general population.

Clinical presentation

Neurological picture. A total of 102 patients reported a neurological symptom. Eighty-six patients (71 females, 41% of female patients, with a mean age of 42 yr ± 1.32; 15 males, 36% of male patients, with a mean age of 40.7 ± 3.32) were first evaluated by the neurologist because of headache and then were referred to us after a sellar CT scan or MRI diag-
nostic for empty sella. Characteristics of headache of these patients were very variable in intensity, quality, course, presentation, and localization. Most of the time, this headache was described as lateral, persistent, and datatable from years. In 21 patients, severe headache was accompanied by symptoms of intracranial hypertension (papilledema and visual disturbances). Rhinorrhea was present in 14 of these patients. In several patients (n = 41), there were other neurological disturbances such as dizziness, syncope, cranial nerve disorders, convulsion, and depression. Two patients had trigeminal neuralgia. Forty-nine patients referred a coexisting endocrine or ophthalmological symptom.

Ophthalmological picture. A total of 41 patients presented with ophthalmological disturbances. Thirty-eight of our patients were first evaluated by the ophthalmologist for a relative worsening of visual acuity, only two of them presented diplopia, one patient had a defect in oculomotor nerve, one had an optical neuritis, and in nine patients a mild papilledema was evidenced.

Endocrine picture and evaluation. In 89 patients (42% of total), an endocrine dysfunction (mainly based on oligomenorrhea in females and sexual dysfunction in males) was initially suspected. In 28 of these patients, coexisting neuroophthalmological symptoms were present. The basal endocrine evaluation (Table 1) demonstrated a tendency toward hyperprolactinemia in the entire female population. In females, basal LH and FSH were higher than in males. This can be explained by increased frequency of frankly hyperprolactinemic subjects among females as well as by the presence of some postmenopausal subjects in the female population. Overall, 40 of 213 patients had documented endocrine abnormalities (19% of the population), specifically 31 females (18% of all females) and nine males (22% of all males).

Hyperprolactinemia. Twenty-two patients (10% of total patients: 18 women, 11% of all women, with a mean age of 38.6 ± 1.1 yr, and four males, with a mean age of 46.5 ± 3.52 yr) presented with hyperprolactinemia. Of these patients, 12 women were in a premenopausal state (7% of all female patients, and 15% of premenopausal women; mean age, 31.5 ± 0.8 yr) and presented with oligomenorrhea, and six were in a postmenopausal state. Six patients had galactorrhoea. All four males referred erectile dysfunction, and two of them presented with gynecomastia. Concerning absolute prolactin levels, almost half of the patients had prolactin between 50 and 100 μg/liter (median prolactin level, 31 μg/liter).

Hypopituitarism. Thirty-six patients underwent pituitary testing for suspected hypopituitarism (Table 2). Anterior hypopituitarism was confirmed in nine patients (six females and three males) according to standard criteria (25). All these patients also had a low fasting cortisol (6.02 ± 1.23 μg/dl), 24-h urinary cortisol (26.73 ± 3.52 μg/24 h), and ACTH (3.46 ± 0.45 pmol/liter). Moreover, eight patients (seven females and one male) presented an isolated GH deficiency. In these patients, plasma IGF-I was below the normal range for sex and age.

In one male subject, studied for remarkable polyuria and polydipsia, central diabetes insipidus was diagnosed with a water deprivation test, suggesting either pituitary stalk or posterior pituitary compression. Subject showed a clinical improvement after DDAVP treatment.

Radiological findings at cerebral MRI/CT scan. Eighty-eight patients underwent a CT scan, and 106 had an MRI (Fig. 3). In 19 patients, both the exams were performed. One hundred thirty-eight of our patients presented a so-called partial empty sella at the CT scan/MRI exam (less than 50% of the sella filled with CSF; pituitary empty sella did not seem to strictly correlate with the severity and/or nature of the clinical picture (7).

<table>
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<th>TABLE 1. Baseline endocrine data in patients with PES</th>
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<td>ACTH</td>
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<td>Cortisol</td>
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<td>24-h urinary free cortisol</td>
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<td>17β-Estradiol</td>
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<td>Testosterone</td>
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Values are expressed as mean ± SEM.
The clinical picture in patients with PES is, therefore, often quite complex and not always is it possible to dissect symptoms and biochemical findings that are the consequences of the empty sella from those casually found that are merely the reason for referral. Therefore, the same symptom may likely be either a direct consequence or casually found in different patients with PES. With this in mind, it is interesting to note that although headache is often the reason that cerebral CT scan or MRI is performed with the finding of PES, and although in a large portion (up to 80%) of subjects with PES, headache (7) has so far been reported, our data show that less than the half of the patients with PES had headache. Clearly, in some patients, the relationship of headache and PES is only casual. However, in other patients with PES, headache may be hypothesized to depend on straining of the suprasellar dural covering (24). Moreover, headache can be part of the clinical picture of intracranial hypertension, although in our study the majority of patients with PES did not have typical signs or symptoms of elevated intracranial pressure (26, 27). Interestingly, based on our experience, other neurological disturbances may have some relevance in the history of patients with PES such as dizziness, syncope, cranial nerve disturbances, and convulsion. More than to a causal relationship with empty sella, this may relate to the increased use of central nervous system imaging in a wide spectrum of neurological diseases.

In 1.6–16% of cases (26, 28, 29), PES has been reported to be associated with visual field defects or other visual abnormalities as a result of posterior dislocation of optic nerve and optic chiasm, optic nerve compression between optic foramen and anterior part of pituitary fossa, partial prolapse of optic tracts into the sella, and optic nerve and optic chiasm straining (30). Our data seem to confirm these previously reported experiences being the incidence of visual problems

<table>
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<tr>
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<th>3 Males (global hypopituitarism)</th>
<th>7 Females (global hypopituitarism)</th>
<th>1 Male (GH deficiency)</th>
<th>7 Females (GH deficiency)</th>
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<tr>
<td><strong>GHRH test (GH peak, ng/liter)</strong></td>
<td>0.27 ± 0.17</td>
<td>0.47 ± 0.27</td>
<td>0.9</td>
<td>0.54 ± 0.24</td>
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<tr>
<td><strong>GnRH test (LH peak, IU/liter)</strong></td>
<td>4.17 ± 2.9</td>
<td>1.58 ± 0.43</td>
<td>18.9</td>
<td>34.47 ± 4.82</td>
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<tr>
<td><strong>GnRH test (FSH peak, IU/liter)</strong></td>
<td>2.0 ± 0.78</td>
<td>2.22 ± 0.55</td>
<td>15.4</td>
<td>27.21 ± 4.71</td>
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<tr>
<td><strong>TRH test (TSH peak, mU/liter)</strong></td>
<td>0.15 ± 0.05</td>
<td>0.17 ± 0.07</td>
<td>14.47</td>
<td>11.61 ± 0.58</td>
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Values are expressed as mean ± SEM.

![Fig. 3. MRI of the empty sella in a representative patient. Coronal (A and B) and sagittal (C and D) weighted T1 images before (A and C) and after (B and D) contrast medium administration. CSF filling the sella turcica with the pituitary stalk remaining in the midline and the pituitary gland flattened against the floor of the sella is reported. oc, Optic chiasm; s, stalk; ca, carotid artery; ss, sphenoid sinus; *, contrast enhancement within the cavernous sinus.](image-url)
in our population of around 15%. The causal relation between these symptoms and PES is reinforced by the observation that visual defects could be an indication for surgical exploration of the empty sella, especially when they are gradually worsening.

In PES particularly, mild hyperprolactinemia has been frequently reported (31). This is thought to be due to pituitary stalk compression as a consequence of the remodeling of the hypothalamo-pituitary region and altered CSF dynamics (32, 33). However, it cannot be excluded that in some patients the association of a high prolactin with PES may be an artifact of submitting patients with elevated prolactin levels to pituitary imaging. Our methodology allowed us, however, to limit this possibility by excluding from the analysis patients with prolactin levels over 100 µg/liter and reducing in the analysis the number of patients with functional hyperprolactinemia by taking into consideration only the prolactin level 40 min after a saline infusion test.

The compression of the pituitary gland and/or the pituitary stalk can cause insufficient function of the gland (34, 35). In the literature (36), global hypopituitarism has been thought to be rare (about 2% of the patients with PES). GH deficiency is apparently the most frequent hormone deficiency described in PES (37–39). In a small number of patients (about 6%), a reduction in gonadotropin release with either pituitary or hypothalamic origin has been observed (39). As well, isolated deficiencies of ACTH, TSH, and antidiuretic hormone have been occasionally described (all about 1% of patients) (40–42).

In our experience, objective endocrine problems were highly represented in the population, accounting overall for about 25% of the patients with PES. Our data are substantially in line with what is reported in the literature. However, true hyperprolactinemia was apparently not so frequent in our patients (10% of total). Reduced frequency with respect to that previously reported may be a result either of the exclusion of secondary empty sella (hyperprolactinemia as a result of pituitary adenoma) or the higher accuracy in evaluating prolactin dynamics. Concerning patients with hypopituitarism, our data confirm that GH deficiency is an emerging feature of patients with PES. This finding is probably related to the nonsystematic quest for this hormonal deficiency (based on IGF-I levels because baseline GH levels are clearly not useful for the diagnosis) (10) and opens the question of whether somatotrophs are a more vulnerable component of the pituitary in PES and of the necessity of doing systematic GH testing in PES also for substitutive treatment purposes. Finally, also in our experience, as previously reported, there is in PES a marginal but not negligible incidence of global hypopituitarism.

Treatment

Results of treatment and follow-up of our patients have been reported, subdividing the population according to if and what type of specific treatment was received. Therefore, we ended up with three categories of patients: medical treatment, surgical treatment, and no treatment.

One hundred nineteen of our patients were submitted to a medical and radiological follow-up; of those patients, 40 were treated with and followed up for specific medical therapy, whereas 21 were treated surgically, and 58 were submitted only to symptomatic medical treatment (defined as untreated).

Results and follow-up of patients submitted to specific medical treatment

Hyperprolactinemia. Of the 22 hyperprolactinemic patients, 17 women and two men were treated pharmacologically because symptomatic (13 with cabergoline and six with bromocriptine), obtaining normalization of circulating prolactin and clinical improvement in all instances. None of these patients underwent surgical treatment. Mean duration of treatment was 3 ± 1 yr. Mean cabergoline dose ranged from 0.5–1.5 mg/wk. No patient was withdrawn from treatment because of side effects.

Hypopituitarism. The nine patients with global anterior hypopituitarism had a complete pituitary hormonal replacement therapy. They had their complete annual follow-up with stable clinical and biochemical substitution. Of the eight patients with isolated adult-onset GH deficiency, only five were submitted to recombinant human GH treatment with no changes in neuroradiological and ophthalmological findings. Those on replacement therapy had a subjective improvement in quality of life, with an increase in strength and wellbeing.

The patient with diabetes insipidus was submitted to DDAVP treatment, which was able to normalize osmolarity (in serum and urine) and serum electrolytes.

Results and follow-up of patients submitted to neurosurgical treatment

The total number of patients submitted to neurosurgery was 21 (17 females and four males). Neurosurgery was performed when signs and symptoms of intracranial hypertension (headache, papilledema, or visual disturbances) of either severe degree or clinically worsening and resistant to medical treatment (acetazolamide, Diamox tab 250 mg, with a dose range of 250–500 mg/d; or escin, Reparil tab 40 mg, with a dose range of 100–250 mg/d) were observed.

In all patients (n = 7) affected by severe intracranial hypertension without rhinorrhea, a CSF shunt device was used. In 14 patients affected by rhinorrhea, 12 patients underwent ventricular-peritoneal shunt and two underwent a primary surgical repair of the sellar floor. The operation stopped rhinorrhea in 10 and reduced it in four of the patients submitted to the shunt. In these last patients, a subsequent surgical repair of sella floor definitively stopped the fistula. Remission of headache, papilledema, and visual disturbances was obtained in all patients.

Results and follow-up of untreated patients

Patients submitted to follow-up not receiving specific treatment were 58 (53 females and five males). Fifty-three of these patients reported originally headache, 12 reported visual alterations, and eight reported oligomenorrhea or sexual dysfunction without demonstrated hormonal alteration. At long-term follow-up (48 ± 7 months), symptomatic medical
treatment caused significant clinical improvement in 48 of 53 patients with headache, six of six patients with oligomenorrhea and two of two patients with sexual dysfunction. Worsening of the symptoms was very infrequent (five patients with headache) as well as appearance of clinical and biochemical alterations in patients originally asymptomatic and with normal hormonal profile (one patient originally euthyroid developed central hypothyroidism years after the diagnosis of empty sella).

Radiological follow-up

More than 50% of the nonoperated patients were submitted to a radiological follow-up with CT scan or MRI (116 of 192). Mean interval of second radiological examination with respect to the diagnosis was 26 ± 7 months. In 85 cases, the radiological degree of empty sella remained unchanged. In 12 patients, a worsening (from partial to total empty sella) of the radiological picture was observed, whereas in 19 patients a partial reversal of the radiological picture was observed.

Discussion

PES may be associated with variable clinical conditions ranging from the occasional discovery of a clinically asymptomatic arachnoid pouch within the sella turcica to severe intracranial hypertension and rhinorrhea (43). The need for replacement hormone therapy in PES, as in other syndromes that may cause hypopituitarism, must be assessed for every single hormone, including GH. The suspicion of symptomatic intracranial hypertension makes CSF shunting procedures necessary to avoid potential serious complications such as blindness.

In our experience, the pharmacological treatment of symptomatic hyperprolactinemia is highly effective both biochemically and clinically. In none of the patients was it possible to suspend dopamine agonist treatment because of the reappearance of symptoms and increase in serum prolactin. Concerning GH substitution in the patients found to have severe GH deficiency, we observed no negative effects but a subjective improvement in the quality of life.

Pharmacological treatment of patients affected by intracranial hypertension, with no or mild to moderate visual loss, has been suggested (44, 45). However, in our patients with increased intracranial pressure, presenting neurological signs (papilledema, severe headache, and visual alterations) or rhinorrhea, only the surgical approach appeared effective in resolving the clinical problem; the placement of a lumbar peritoneal shunt was a satisfactory treatment for the majority of the patients with increased intracranial pressure, although during follow-up some patients required multiple shunt revisions. Occasionally, transsphenoidal surgical repair of the sellar defect may be necessary. Optic nerve sheath decompression has also been advocated as a method to alleviate symptoms and signs, such as papilledema, although this technique fails to directly address the issue of elevated intracranial pressure and may have a nonnegligible failure rate (46–49).

In conclusion, patients with PES should always be submitted to endocrine, neurological, and ophthalmological evaluation at presentation because of the very high incidence of these abnormalities. Endocrine screening should be followed by specific endocrine testing when hormonal abnormalities are suspected. This procedure is able to detect all the patients with a very low number of unconfirmed diagnoses.

All endocrine deficits should be treated with appropriate medical substitution. Hyperprolactinemia always improves after dopamine agonist treatment. In those patients with PES accompanied by signs and symptoms of intracranial hypertension, severe headache, visual alterations, and rhinorrhea, surgery is necessary.

Patients with no abnormalities at baseline are unlikely to develop neuro-ophthalmological symptoms or endocrine abnormalities in the follow-up. Moreover, also the radiological degree of PES tends to remain constant over time. However, because of the theoretical risk of progression, a reevaluation after 24–36 months (if there are not clinical indications before) of the endocrine, neuro-ophthalmological and radiological picture is reasonable. If progression is not observed, additional control evaluation could be even less frequent and limited to those patients requiring it clinically. Patients treated medically for any endocrine abnormalities should be reevaluated according to appropriate well-established guidelines. Patients treated surgically should also be reevaluated for assessing long-term results and side effects at least twice in the year after surgery.

Acknowledgments

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