e-ISSN: 2747-2051

Archives of the Medicine and Case Reports

[AMCR]

https://hmpublisher.com/index.php/amcr

Reversible Movement Disorder Unmasking Secondary Fahr Syndrome after Total Thyroidectomy: A Case of Bilateral Basal Ganglia and Cerebellar Calcification from Chronic Hypoparathyroidism

Made Ngurah Jiyesta Wibawa[1*], I Putu Ananta Wijaya Sabudi[2]

[1]General Practitioner, Klungkung Regional General Hospital, Semarapura, Indonesia

[2]Neurosurgeon, Klungkung Regional General Hospital, Semarapura, Indonesia

ARTICLE INFO

Keywords:
Basal ganglia calcification
Fahr syndrome
Hypocalcaemia
Hypoparathyroidism
Movement disorders

***Corresponding author:**
Made Ngurah Jiyesta Wibawa

E-mail address:
jiyestangurah@gmail.com

All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.37275/amcr.v7i3.918

A B S T R A C T

Fahr syndrome denotes bilateral, symmetrical calcification of the basal ganglia and other deep brain structures occurring secondary to an identifiable disorder, most often hypoparathyroidism, and must be distinguished from idiopathic Fahr disease. Clinical data from Indonesia remain limited, and the movement disorder that heralds the condition is frequently mistaken for primary neurological disease. We report a 35-year-old woman who presented with a one-hour history of involuntary movements of the face and all four limbs and dysarthria from excessive facial muscle contraction, superimposed on a seven-year history of intermittent right-hand paraesthesia. Her background included a total thyroidectomy seven years earlier with subsequent levothyroxine-treated hypothyroidism, chronic hypocalcaemia presumed to reflect post-surgical hypoparathyroidism (calcium carbonate 500 mg every 12 hours), and a six-year diagnosis of "idiopathic" generalised epilepsy made without neuroimaging. Examination showed full consciousness, a positive Chvostek sign, mild four-limb ataxia and minor memory impairment. Non-contrast cranial computed tomography demonstrated multiple calcifications in both parietal lobes, the basal ganglia and both cerebellar hemispheres, without haemorrhage or infection. Serum calcium was 4.5 mg/dL and parathyroid hormone was below 6 pg/mL, confirming secondary Fahr syndrome from post-thyroidectomy hypoparathyroidism. Treatment with calcium carbonate, phenytoin, levothyroxine and vitamin B complex produced complete resolution of the involuntary movements and seizures and marked improvement of ataxia within six weeks, sustained at two months. This case underscores the need to screen phosphocalcic metabolism and obtain neuroimaging before labelling epilepsy idiopathic in any thyroidectomised patient, and the value of sustained metabolic control as both treatment and secondary prevention.

1. Introduction

Fahr syndrome is a rare neuro-metabolic disorder defined by bilateral, symmetrical calcification of the basal ganglia and other deep grey and white matter structures, including the thalamus, dentate nucleus, cerebral cortex and subcortical white matter, arising as a consequence of an identifiable underlying disease.[1,2] The calcification is frequently detected incidentally on neuroimaging, both in asymptomatic individuals

and in patients who already display neurological or psychiatric symptoms, and its recognition should prompt a search for an underlying cause rather than being dismissed as an isolated finding.[1,2] The eponym is often used loosely, but a precise distinction carries clinical weight. Fahr disease — increasingly termed primary familial brain calcification — describes idiopathic or hereditary calcification occurring in the absence of any metabolic derangement, and has been linked to autosomal-dominant mutations in *SLC20A2*, *PDGFRB*, *PDGFB* and *XPR1*, and to autosomal-recessive mutations in *MYORG*.[3,4] Fahr syndrome, by contrast, denotes the secondary form, in which the calcification is driven by a definable cause, most commonly a disorder of parathyroid hormone (PTH) and calcium–phosphate homeostasis.[1,5] Correctly assigning a patient to the syndrome rather than the disease reframes the illness as potentially treatable, because control of the underlying metabolic lesion can arrest and sometimes reverse the neurological manifestations.[1,2]

The reported prevalence of brain calcification meeting the criteria for Fahr disease or syndrome is under one per million in the general population, yet the frequency of basal ganglia calcification among patients with chronic hypoparathyroidism is far higher, with series describing rates from roughly 12% to 74%.[6,7] In a systematic review of 223 published cases of hypoparathyroid Fahr syndrome, the mean age at presentation was approximately 44 years with a female predominance; about 39% of cases were idiopathic, 35% acquired (chiefly post-surgical) and 26% due to pseudohypoparathyroidism.[5] Almost half of the patients in that review had tetany, seizures or a movement disorder, and approximately 40% displayed neuropsychiatric symptoms, illustrating that the condition, though individually rare, produces a heavy and heterogeneous symptom burden when it does occur.[5] Hypoparathyroidism — whether idiopathic, autoimmune or, most often, acquired after anterior neck surgery — produces sustained hypocalcaemia and hyperphosphataemia that promote abnormal mineral deposition within the brain.[6,8] The severity and, above all, the duration of hypocalcaemia are considered the principal determinants of intracerebral calcification.[7,9] Calcium homeostasis depends on the tightly coordinated action of PTH, vitamin D and the target organs of bone, kidney and gut. Parathyroid hormone raises the plasma calcium concentration by mobilising skeletal calcium, enhancing distal renal tubular calcium reabsorption while promoting phosphaturia, and stimulating renal 1α-hydroxylase to generate calcitriol, which in turn augments intestinal calcium absorption.[6,7] When the parathyroid glands are removed or devascularised — the commonest scenario being inadvertent injury during thyroid surgery — this regulatory axis collapses, producing the biochemical signature of hypoparathyroidism: hypocalcaemia, hyperphosphataemia and inappropriately low or undetectable PTH.[8,10] Post-surgical hypoparathyroidism is the single most frequent cause of chronic hypoparathyroidism, and although the majority of cases that follow thyroidectomy are transient, a minority become permanent and expose the patient to years of metabolic disturbance and its long-term complications, of which intracerebral calcification is among the most striking.[8,10]

Clinically, Fahr syndrome is strikingly protean. Neurological manifestations include seizures and extrapyramidal movement disorders such as dystonia, parkinsonism, tremor and choreoathetosis, together with cerebellar ataxia; cognitive features range from mild memory

impairment to progressive dementia, and psychiatric features encompass depression, anxiety, hallucinations and frank psychosis.[5,11,12] The specific symptom profile in an individual patient is thought to reflect the topography of calcification, so that predominant basal-ganglia involvement produces movement disorders, cerebellar deposits produce ataxia and dysarthria, and cortical or hippocampal deposits contribute to cognitive and psychiatric features.[2,13] Patients with hypoparathyroidism are additionally exposed to the consequences of acute hypocalcaemia — neuromuscular irritability with a positive Chvostek or Trousseau sign, tetany, laryngospasm and seizures that can escalate to status epilepticus — which raise morbidity and occasionally mortality and demand urgent correction.[14,15] Because these features so closely resemble primary epileptic, movement and psychiatric disorders, diagnosis is frequently delayed, particularly where access to neuroimaging is limited, and patients may be treated for years for a presumed primary neurological illness before the underlying metabolic lesion is uncovered.[10,11] Despite a well-established association between hypoparathyroidism and Fahr syndrome, important gaps remain, especially in Indonesia, where documented cases are few.[10] No therapy is yet able to dissolve established intracerebral calcification; management remains directed at correcting hypocalcaemia and controlling neurological symptoms.[1,16] The absence of standardised, universally accepted diagnostic criteria contributes to inconsistent recognition and reporting, particularly in resource-limited settings, and epidemiological data on the incidence and natural history of the secondary form in hypoparathyroid populations are sparse, hampering the development of early-screening protocols and evidence-based management pathways.[5,7] Furthermore, because the calcification distribution and clinical severity vary widely between individuals — some patients remaining asymptomatic for decades while others develop disabling movement disorders — the factors that determine who progresses to symptomatic disease are poorly defined.[2,7] Reports that carefully document the metabolic, radiological and therapeutic course therefore retain considerable educational value.

The diagnostic approach to suspected Fahr syndrome rests on the convergence of three lines of evidence: a compatible clinical syndrome, characteristic bilateral symmetrical calcification on non-contrast CT, and biochemical confirmation of the underlying metabolic disorder — in the majority of cases, hypocalcaemia with an inappropriately low PTH.[1,2] In practice, the first opportunity to make the diagnosis is often missed: patients present to general practitioners, neurologists or psychiatrists with seizures, movement disorders or behavioural change, and unless calcium metabolism is checked and a head CT obtained, the metabolic origin can remain hidden for years.[10,11] This diagnostic latency is precisely what unfolded in the patient described here, and it frames the practical lessons that this report seeks to convey. The novelty of the present report is threefold. First, it documents an unusually *widespread* calcification distribution — extending beyond the basal ganglia to the parietal cortex and to both cerebellar hemispheres — in a comparatively young woman whose hypoparathyroidism was post-surgical, a mechanism generally associated with a *lower* calcification burden than non-surgical disease.[7] Second, it illustrates how a longstanding diagnosis of “idiopathic” generalised epilepsy, made years earlier without any neuroimaging, obscured a treatable metabolic cause. Third, it adds much-needed Indonesian clinical data to a literature

dominated by reports from other regions.[1,10] The aim of this case report is to describe the clinical, radiological and biochemical features of secondary Fahr syndrome due to post-thyroidectomy hypoparathyroidism, and to discuss a diagnostic and management approach that may accelerate recognition and improve outcomes in similar patients.

2. Case Presentation

A 35-year-old woman presented to the neurosurgery outpatient clinic of Klungkung Regional General Hospital with involuntary movements of the face, upper limbs and lower limbs that had begun one hour before arrival. The movements had appeared abruptly, were most prominent when she was fatigued, and could not be voluntarily suppressed. She also reported difficulty speaking (dysarthria) attributable to excessive contraction of the facial muscles. The involuntary movements were described as sporadic, with intensity that increased with tiredness. In addition, she had experienced intermittent tingling (paraesthesia) of the right hand over the preceding seven years, a symptom that had begun after she underwent total thyroidectomy for the treatment of thyroid disease. She denied seizures, nausea, vomiting, severe headache or any decrease in consciousness since the onset of the involuntary movements.

Her medical history was significant for idiopathic generalised epilepsy diagnosed six years earlier — notably, without any brain imaging having been performed at the time — for which she had taken phenytoin, clobazam and folic acid regularly for the preceding six years. Seven years earlier she had been diagnosed with hypothyroidism after undergoing total thyroidectomy, and she remained on levothyroxine. She had also been diagnosed with hypocalcaemia seven years previously, presumed to result from post-thyroidectomy hypoparathyroidism, and had been taking oral calcium supplementation (calcium carbonate 500 mg every 12 hours) since then. She had no history of stroke, diabetes mellitus, hypertension, heart disease, hypercholesterolaemia or malignancy. There was no family history of similar neuromuscular complaints, and she denied any use of alcohol, tobacco or illicit drugs. Of particular note, no brain imaging or dedicated assessment of calcium metabolism had been undertaken at the time her epilepsy was first diagnosed, so the relationship between her chronic hypocalcaemia and her seizure disorder had never been examined. This convergence of a post-surgical endocrine history, chronic hypocalcaemia and an unexplained seizure disorder — each managed in isolation — provided the essential background against which her acute presentation was to be interpreted.

On physical examination the patient was fully conscious, with a Glasgow Coma Scale score of E4V5M6 (15). Her vital signs and principal examination findings are detailed in Table 1. Blood pressure was 126/86 mmHg, heart rate 106 beats per minute, respiratory rate 20 breaths per minute, axillary temperature 36.0 °C and oxygen saturation 97% on room air. Neurological examination revealed minor memory impairment and a positive Chvostek sign, elicited as contraction of the facial muscles over the frontal, periorbital and cheek regions following percussion. Mild ataxia was present in all four extremities, pointing towards bilateral cerebellar dysfunction. The involuntary movements affected the face and all four limbs and were accompanied by tremor and dystonic posturing that fluctuated in intensity, being most pronounced when the patient was fatigued. Cranial-nerve examination was otherwise intact

aside from the dysarthria, muscle power and tone were preserved between episodes of involuntary movement, and there were no meningeal signs. The remainder of the general and systemic examination was unremarkable.

Table 1. Vital signs and neurological examination findings on admission.

Parameter	Finding	Reference / interpretation
Glasgow Coma Scale	E4V5M6 (15)	Normal — fully conscious
Blood pressure	126/86 mmHg	Normal
Heart rate	**106 beats/min**	Mild sinus tachycardia
Respiratory rate	20 breaths/min	Normal
Axillary temperature	36.0 °C	Normal
Oxygen saturation	97% on room air	Normal
Chvostek sign	**Positive**	Neuromuscular irritability (hypocalcaemia)
Limb coordination	**Mild ataxia, four limbs**	Suggests bilateral cerebellar dysfunction
Cognition	**Minor memory impairment**	Consistent with cortical/subcortical calcification
Involuntary movements	**Face and all four limbs**	Extrapyramidal manifestation
Speech	**Dysarthria**	From excessive facial-muscle contraction

Notes: Abnormal values are shown in bold red.

Non-contrast computed tomography (CT) of the head demonstrated multiple calcifications in the right and left parietal regions, in the basal ganglia bilaterally, and in the right and left cerebellar hemispheres, with no evidence of intracranial haemorrhage or cerebral infection (Figure 1). The bilateral, symmetrical distribution of the calcific deposits, involving both supratentorial and infratentorial structures, was highly characteristic of Fahr syndrome. As shown in Figure 1A, dense calcific foci were evident within the basal ganglia at the level of the lateral ventricles, while Figure 1B demonstrates calcification within the posterior fossa at the level of the cerebellar hemispheres.

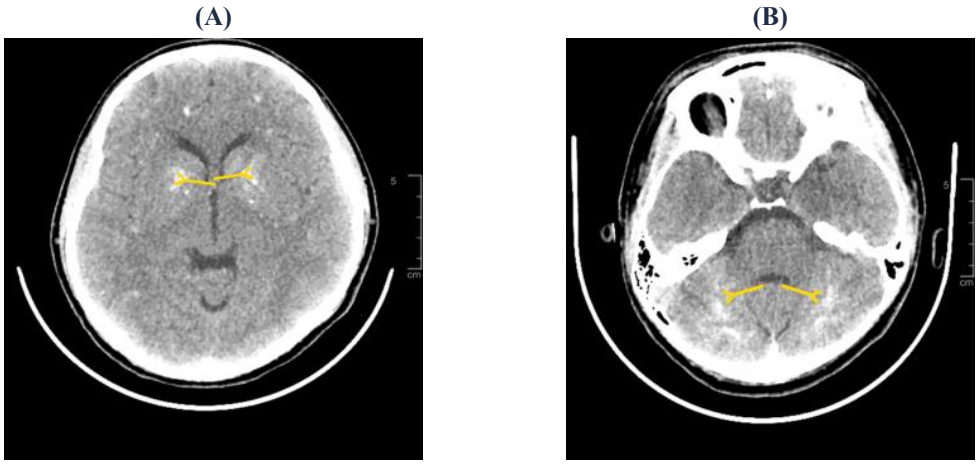

Figure 1. Non-contrast axial cranial CT. (A) At the level of the lateral ventricles, symmetrical hyperdense calcific foci are present within the basal ganglia bilaterally (arrows), with additional scattered parietal calcification. (B) At the posterior-fossa level, calcification is seen within both cerebellar hemispheres (arrows). No intracranial haemorrhage or infective focus is identified. Patient identifiers have been removed.

Initial laboratory investigation, summarised in Table 2, revealed a parathyroid hormone (PTH) concentration below 6 pg/mL and a markedly reduced serum calcium of 4.5 mg/dL, confirming severe hypocalcaemia in the context of inappropriately low PTH. The finding of hypocalcaemia together with a suppressed PTH is the defining biochemical hallmark of hypoparathyroidism and, in this patient, provided the metabolic explanation for both the acute neuromuscular signs and the chronic intracerebral calcification.[6,7] Integrating the clinical, radiological and biochemical findings, the patient was diagnosed with Fahr syndrome secondary to hypoparathyroidism, chronic hypocalcaemia following total thyroidectomy, and idiopathic generalised epilepsy.

Table 2. Initial laboratory results.

Test	Result	Reference range	Interpretation
Parathyroid hormone (PTH)	**< 6 pg/mL**	15–65 pg/mL	**Inappropriately low**
Serum calcium (total)	**4.5 mg/dL**	8.6–10.2 mg/dL	**Severe hypocalcaemia**

Notes: Abnormal values are shown in bold red. Serum phosphate, magnesium and vitamin D were not available for this patient.

The diagnostic reasoning proceeded by pattern recognition and exclusion. The acute hyperkinetic movement disorder in a young adult raised a broad differential that included primary movement disorders, drug-induced dyskinesia, autoimmune and metabolic encephalopathies, and secondary brain calcification syndromes. The combination of a positive Chvostek sign, longstanding paraesthesia and a background of thyroid surgery immediately directed attention to a disorder of calcium metabolism, and the bilateral, symmetrical calcification on CT — involving the basal ganglia, parietal cortex and cerebellum without haemorrhage, mass effect or infective features — pointed firmly towards Fahr syndrome rather than an alternative structural lesion.[1,2] The absence of a family history of similar disease, of syndromic dysmorphism, and of any relevant toxic or infective exposure argued against primary familial brain calcification and against the genetic, infectious and toxic causes of secondary calcification, while the documented biochemical picture confirmed hypoparathyroidism as the operative mechanism.[4,17,18] Her earlier label of “idiopathic” generalised epilepsy was, in retrospect, most plausibly an early hypocalcaemic manifestation of the same disorder that had never been imaged.

Management, detailed in Table 3, comprised vitamin B complex every 12 hours for neurotrophic support, phenytoin 200 mg every 12 hours for control of epilepsy, levothyroxine 150 µg every 24 hours for the management of hypothyroidism, and calcium carbonate 500 mg every 12 hours for correction of hypocalcaemia. She was then followed as an outpatient with close monitoring for further evaluation of her neurological symptoms and adjustment of her metabolic therapy. The treatment strategy deliberately combined correction of the underlying metabolic disorder with symptomatic control: calcium carbonate addressed the hypocalcaemia that lay at the root of both the neuromuscular irritability and the intracerebral

calcification, phenytoin controlled the seizure tendency to which chronic hypocalcaemia predisposes, levothyroxine maintained euthyroidism after her earlier thyroidectomy, and neurotrophic support was added as an adjunct. Physical activity and gentle motor rehabilitation were encouraged to support recovery of coordination and balance.

Table 3. Pharmacological management regimen initiated at diagnosis.

Agent	Dose and route	Indication
Calcium carbonate	500 mg orally every 12 h	Correction of hypocalcaemia
Phenytoin	200 mg orally every 12 h	Control of epilepsy / seizure prophylaxis
Levothyroxine	150 µg orally every 24 h	Post-thyroidectomy hypothyroidism
Vitamin B complex	Orally every 12 h	Neurotrophic support

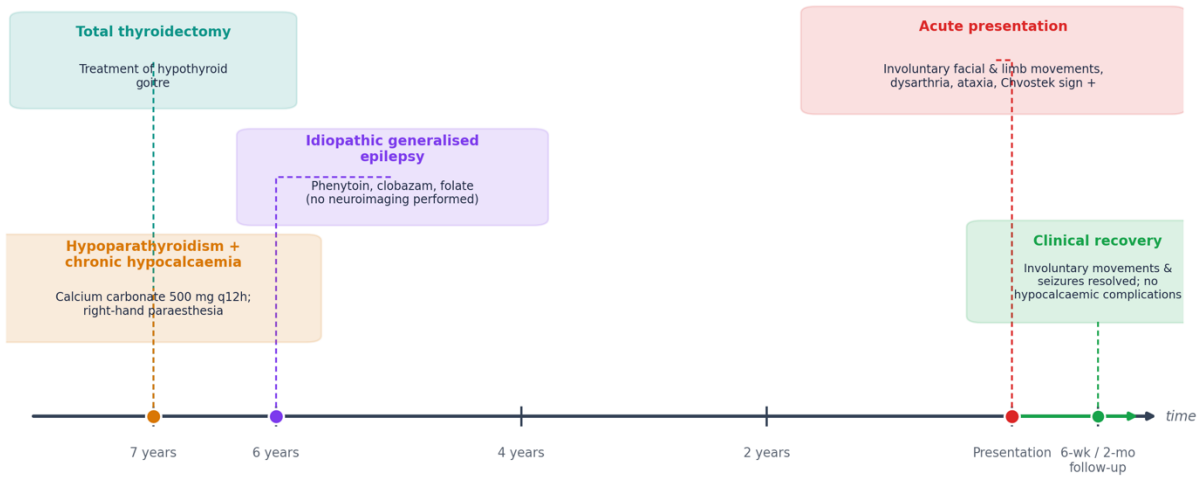

Figure 2. Seven-year clinical course. Total thyroidectomy was followed by hypothyroidism and chronic hypoparathyroid hypocalcaemia; an "idiopathic" generalised epilepsy was diagnosed one year later without neuroimaging. The acute presentation with a movement disorder led to the diagnosis of secondary Fahr syndrome, with clinical recovery after metabolic correction and anticonvulsant optimisation.

After six weeks of therapy the patient showed a favourable outcome: the involuntary movements of the face and extremities resolved completely, tremor and dystonia were drastically reduced, and the mild ataxia subsided. Clinically she experienced no further seizure episodes and was able to resume her daily activities without significant neurological limitation. Over a two-month follow-up period there were no complications of hypocalcaemia and no adverse effects attributable to the anticonvulsant, so the same therapeutic regimen was continued and routine monitoring was recommended. The overall course, from thyroidectomy to clinical recovery, is summarised in Figure 2, and the trajectory of her neurological symptom burden before and after treatment is depicted in Figure 3.

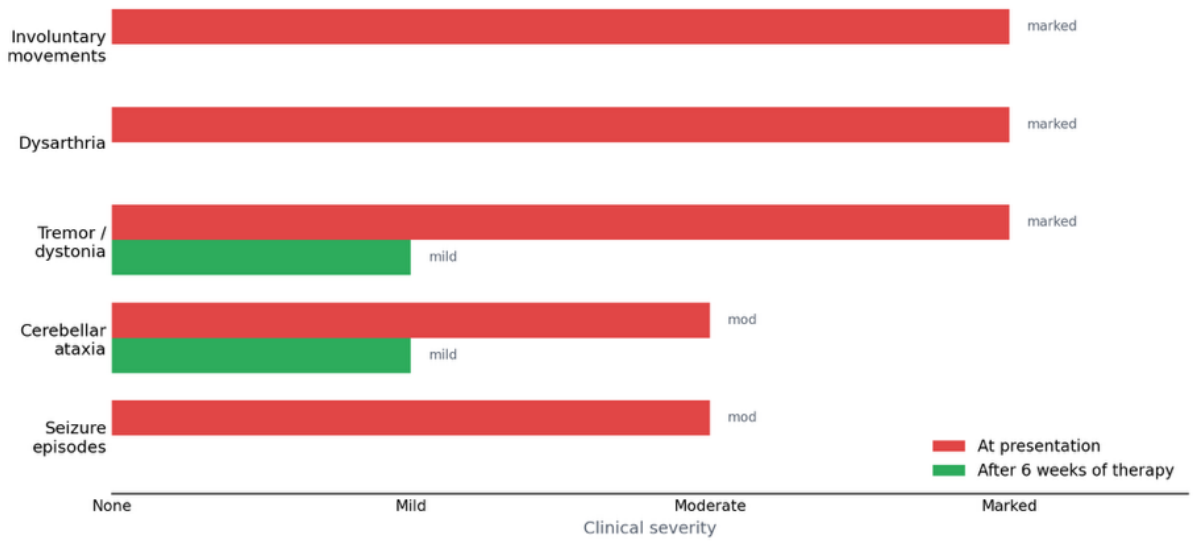

Figure 3. Neurological symptom burden at presentation compared with six weeks after treatment. Involuntary movements, dysarthria and seizures resolved completely, while tremor/dystonia and cerebellar ataxia were reduced to mild residual severity. No hypocalcaemic or drug-related complications occurred during two months of follow-up.

3. Discussion

Fahr syndrome, also known as bilateral strio-pallido-dentate calcinosis, is a rare neurological disorder characterised by abnormal calcium deposition in several brain regions, particularly the basal ganglia, thalamus, dentate nucleus, hippocampus, cerebral cortex and subcortical cerebellar white matter.[1,2] The condition may be idiopathic or secondary to another disorder, such as hypoparathyroidism, which disrupts calcium and phosphate metabolism.[1,6] In the present patient, the combination of documented post-thyroidectomy hypoparathyroidism, severe hypocalcaemia and inappropriately low PTH firmly places the illness within the *secondary* category — Fahr syndrome rather than Fahr disease — a distinction that is not merely semantic, because it identifies a modifiable metabolic driver and thereby a treatment target.[1,2]

The clinical picture of Fahr syndrome is highly variable and depends on the location and extent of calcification. Movement disorders such as dystonia, tremor, myoclonus and parkinsonism are common, and cognitive and psychiatric disturbances, including dementia, hallucinations and behavioural change, may also occur.[11,12] In this patient the involuntary movements of the face, hands and feet, together with dysarthria, correspond to the extrapyramidal manifestations of the syndrome, while the mild four-limb ataxia is consistent with the bilateral cerebellar calcification seen on CT. Her longstanding history of “idiopathic” generalised epilepsy is highly relevant: seizures are among the most frequently reported neurological features of hypoparathyroid Fahr syndrome — sometimes the prominent presenting symptom — and chronic hypocalcaemia is itself epileptogenic.[11,14,15,19] It is therefore probable that her seizure disorder, diagnosed six years earlier without neuroimaging, was in fact an early manifestation of the same

metabolic process, misclassified in the absence of a CT scan and biochemical screening. The systematic review by Kalampokini and colleagues quantified this symptom clustering, reporting that patients with a movement disorder carried more than twice the odds of concurrent neuropsychiatric symptoms and that phenotype severity correlated significantly with the extent of brain calcification.[5]

Distinguishing the secondary syndrome from primary familial brain calcification also has prognostic and counselling implications, and warranted deliberate exclusion in this patient. Primary familial brain calcification is genetically heterogeneous: autosomal-dominant forms arise from mutations in *SLC20A2* and *XPR1*, which encode inorganic-phosphate transporters, and in *PDGFB* and *PDGFRB*, which maintain the integrity of the neurovascular unit, whereas the autosomal-recessive form is caused by biallelic *MYORG* variants and tends to produce prominent cerebellar signs and earlier, more severe calcification.[3,4] These monogenic disorders share the radiological phenotype of Fahr syndrome but occur, by definition, with normal calcium and PTH; the presence of unequivocal hypoparathyroid biochemistry in our patient, together with the lack of any family history, made a primary genetic cause improbable and obviated the need for genetic testing in the acute setting.[4,18] The breadth of alternative secondary causes — genetic syndromes such as the 22q11.2 deletion (DiGeorge) syndrome and mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes (MELAS), as well as infectious, inflammatory and toxic aetiologies — is summarised in Table 4, and each was considered and excluded on clinical, biochemical or radiological grounds.[13,17,20]

The CT findings in this patient — multiple calcifications in the parietal lobes, basal ganglia and both cerebellar hemispheres — represent the imaging hallmark of Fahr syndrome, and non-contrast CT is widely regarded as the most valuable and accessible modality for its detection.[1,2] These appearances mirror numerous reports of symmetrical basal ganglia calcification arising from hypoparathyroidism, including idiopathic and post-surgical forms in patients presenting with recurrent involuntary movements, a positive Chvostek sign, hypocalcaemia, hyperphosphataemia and low PTH.[6,21,22] Of particular relevance is the Indonesian report by Supit and colleagues, who described a 35-year-old woman with post-thyroidectomy hypoparathyroidism, a serum calcium of approximately 4 mg/dL, seizures and extensive bilateral basal ganglia calcification — an almost exact demographic and mechanistic counterpart to the present case, and a reminder that this presentation is being encountered, and increasingly recognised, in the Indonesian setting.[10] The additional cortical and cerebellar involvement in our patient argues that her calcification was unusually widespread, consistent with the prolonged and severe nature of her hypocalcaemia.[7]

From an imaging standpoint, non-contrast CT remains the investigation of choice for detecting and characterising intracerebral calcification because of its high sensitivity to mineral density, its wide availability and its low cost — attributes that are especially important in resource-constrained settings such as the one in which this patient was managed.[1,2] Magnetic resonance imaging is complementary rather than confirmatory for calcification; although it delineates associated parenchymal changes and can display calcium with variable signal on gradient-echo and susceptibility-weighted sequences, it is less specific than CT for the mineral itself. Functional dopaminergic imaging with a dopamine-transporter (DaT) scan has been

explored in patients presenting with parkinsonism, and reported cases have shown that extensive basal-ganglia calcification may coexist with a normal DaT scan, implying that the movement disorder does not always reflect nigrostriatal dopaminergic denervation and that its mechanism is more complex than simple neuronal loss.[5] In our patient, the characteristic CT appearance in the appropriate biochemical context was sufficient for diagnosis, and advanced imaging was not required.

Although the precise mechanism of calcium deposition remains incompletely understood, several lines of evidence implicate dysregulated calcium and phosphate metabolism, chiefly driven by hypoparathyroidism, in ectopic mineralisation of brain tissue. Parathyroid hormone maintains the plasma calcium concentration through several actions: it increases renal calcium reabsorption, stimulates the release of calcium from bone, and enhances the production of calcitriol, which augments intestinal calcium absorption.[6,7] In hypoparathyroidism, deficient PTH secretion produces hypocalcaemia and hyperphosphataemia through impaired renal calcium reabsorption and reduced phosphate excretion, while the associated fall in active vitamin D further diminishes intestinal calcium absorption.[6,16] The resulting chronic hypocalcaemia and phosphate excess create a biochemical environment that favours pathological calcium–phosphate precipitation, as summarised in Figure 4.

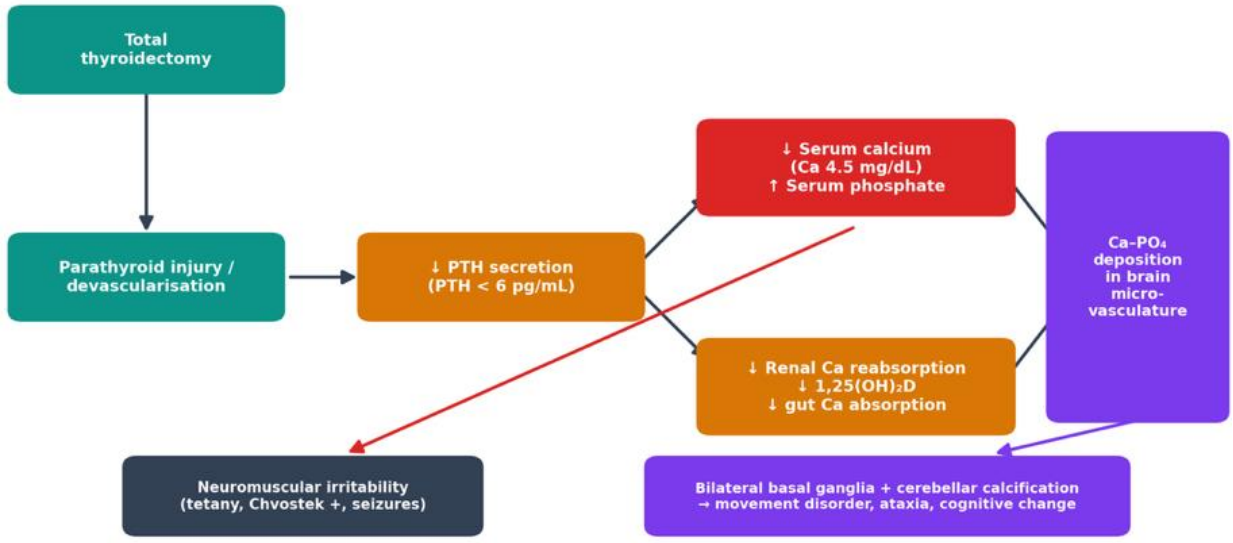

Figure 4. Proposed pathophysiological cascade. Parathyroid injury during total thyroidectomy reduces PTH secretion, producing hypocalcaemia and hyperphosphataemia through diminished renal calcium reabsorption, reduced 1,25-dihydroxyvitamin D and decreased intestinal calcium absorption. The resulting calcium–phosphate excess promotes deposition within the cerebral microvasculature, yielding bilateral basal-ganglia and cerebellar calcification, while acute hypocalcaemia drives neuromuscular irritability, tetany and seizures.

Several complementary mechanisms have been proposed to translate this metabolic disturbance into localised brain calcification. An elevated phosphate concentration within the cerebrospinal fluid may disturb the permeability of the blood–brain barrier, allowing excess calcium and phosphate to be deposited in cerebral tissue, preferentially in metabolically active regions such as the basal ganglia and dentate nucleus.[2,23] Experimental striatal-cell models have shown that a combination of altered 1,25-dihydroxyvitamin D, calcium and even moderately elevated phosphate up-regulates pro-osteogenic molecules — including Wnt3a, alkaline phosphatase, collagen-1α and neuronal phosphate transporters — that actively drive a bone-like mineralisation programme, and, importantly, that conventional calcitriol and calcium therapy can in some circumstances enhance rather than suppress this process.[9] Chronic hypocalcaemia has additionally been linked to mitochondrial dysfunction, increased free-radical production and endothelial injury within the cerebral microvasculature, which may accelerate calcification in chronically hypoperfused regions.[9,23] A detailed neuropathological study confirmed that the calcium deposits lie predominantly within the walls of arterioles and capillaries and extend along penetrating medullary arteries, establishing the microvascular substrate of the disorder.[23]

The large clinical study by Zavatta and colleagues provides the most robust quantitative framework for interpreting our patient's imaging. In a 20-year case-control analysis of 142 adults with chronic hypoparathyroidism, basal ganglia calcification was present in about a quarter of patients, appeared at a younger age than in controls, and — counterintuitively — was more than five-fold more common in non-surgical than in post-surgical disease.[7] Crucially, calcification correlated with low serum calcium and a low calcium/phosphate ratio, but not with phosphate alone or the calcium–phosphate product, and lower serum calcium was associated with a greater volume of calcification.[7] Viewed against this background, our patient is instructive: despite a post-surgical mechanism generally associated with a lower calcification burden, she developed extensive supratentorial and infratentorial deposits, underscoring that the depth and chronicity of her hypocalcaemia — a calcium of 4.5 mg/dL sustained over seven years — were sufficient to override the relative protection conferred by the surgical aetiology. The breadth of causes that can produce an identical radiological picture is summarised in Table 4, ranging from primary familial brain calcification through endocrine, genetic, infectious and toxic aetiologies.[4,13,17,20]

Placing this case within the published literature reinforces both its typicality and its distinctive features. Case series from different regions have repeatedly documented the triad of hypocalcaemia, low PTH and symmetrical basal-ganglia calcification: Berrabeh and colleagues described five patients in whom neurological and cognitive disorders, seizures and abnormal movements associated with tetany led to the recognition of hypoparathyroid Fahr syndrome, all of whom improved with treatment; Arruda and colleagues reported four further cases and emphasised that early treatment may prevent calcification and neurological deterioration.[1,6] Individual reports have highlighted particular precipitants and presentations — status epilepticus following infection, psychotic features accompanying hypocalcaemia, parkinsonism in post-surgical disease, and hypocalcaemic seizures requiring intensive care — underlining the phenotypic range

of the disorder.[11,14,15,21] Against this backdrop, our patient is characteristic in her biochemistry and imaging but notable for the combination of a relatively young age, a post-surgical mechanism and an unusually extensive calcification distribution, and for the clarity with which her earlier "idiopathic" epilepsy can be reinterpreted as an unrecognised hypocalcaemic manifestation.[5,10]

Table 4. Principal causes of bilateral brain calcification and their relevance to the present patient.

Category	Representative causes	Relevance to this patient
Primary (Fahr disease)	Familial / idiopathic; SLC20A2, PDGFB, PDGFRB, XPR1, MYORG mutations	Excluded — a clear metabolic cause was present; no family history
Endocrine / metabolic	Hypoparathyroidism (idiopathic, post-surgical, autoimmune); pseudo- and pseudopseudohypoparathyroidism; hyperparathyroidism	**Confirmed cause — post-thyroidectomy hypoparathyroidism (PTH < 6 pg/mL, Ca 4.5 mg/dL)**
Genetic syndromes	DiGeorge (22q11.2 deletion); MELAS; Cockayne syndrome	Unlikely — no syndromic or mitochondrial features
Infectious / inflammatory	Congenital TORCH infections; neurocysticercosis; brucellosis	Excluded — no infective focus on CT or clinically
Toxic / physical	Lead or carbon-monoxide exposure; prior cranial irradiation	Excluded — no relevant exposure history

The way in which basal-ganglia calcification generates a movement disorder merits specific consideration, because it is not simply a matter of physical mineral bulk. The basal ganglia form part of cortico-striato-pallido-thalamo-cortical loops that regulate the initiation, scaling and suppression of movement, and calcium deposition within the striatum, globus pallidus and their microvasculature is thought to disrupt these circuits through a combination of local neuronal dysfunction, impaired neurotransmission and chronic microvascular ischaemia.[2,23] This model helps to explain why the clinical severity correlates with the extent, rather than merely the presence, of calcification, and why some patients with dopaminergic features nonetheless retain a normal dopamine-transporter scan.[5] It also accounts for the mixed hyperkinetic phenotype seen in our patient, in whom involuntary movements, tremor and dystonia coexisted with cerebellar ataxia, reflecting simultaneous supratentorial and infratentorial involvement. Importantly, because the underlying neuronal architecture is disrupted rather than destroyed in the earlier stages, restoration of a normal metabolic milieu can allow substantial functional recovery, as was observed here.[1,2]

No definitive treatment currently exists to remove established brain calcification, and management is therefore directed at controlling symptoms and correcting the underlying metabolic derangement.[1,2] Across the published case reports and series, the consistent first-line approach is conventional therapy with calcium — given intravenously in the acute, severely hypocalcaemic setting and orally for maintenance — together with an active vitamin D metabolite such as calcitriol, alongside anticonvulsant therapy where seizures are present.[8,12,16,24] Reported outcomes are encouraging: correction of hypocalcaemia with calcium and calcitriol has been described to

reverse symptoms and normalise biochemistry, and cognitive and motor features frequently improve or stabilise once the metabolic disturbance is addressed.[1,16,21] In our patient, treatment combined calcium carbonate to address the hypocalcaemia, phenytoin for seizure control, levothyroxine for hypothyroidism and vitamin B complex for neurotrophic support; sustained long-term control of serum calcium, ideally kept within the low-normal range with regular biochemical monitoring while avoiding over-correction, is the shared lesson of these reports and the rationale for continued outpatient surveillance.[7,16] The involuntary movements and seizures resolved and the ataxia improved markedly within six weeks, echoing the favourable responses reported across multiple case series once hypocalcaemia is corrected and anticonvulsant therapy is optimised.[1,21,22] The importance of sustained biochemical control is amplified by the experimental observation that supraphysiological phosphate exposure promotes further calcification, arguing for meticulous, long-term normalisation of calcium and phosphate rather than intermittent correction.[7,9]

The prognosis of Fahr syndrome is variable and depends on the severity of symptoms and the response to treatment. Early identification and appropriate management of hypocalcaemia can halt or even partially reverse neuropsychiatric manifestations, and cognitive stabilisation has been described once calcium levels are normalised.[1,11] Conversely, extensive established calcification, particularly when accompanied by prolonged hypocalcaemia, may result in permanent neurological deficits such as treatment-resistant parkinsonism and irreversible cognitive decline, while systemic manifestations of hypoparathyroidism — extracranial vascular and renal calcification, nephrolithiasis and cataract — add to long-term morbidity.[5,16] The near-complete recovery achieved in our patient reinforces the value of prompt recognition and metabolic correction, but her wide calcification burden also signals a need for continued vigilance.

Notwithstanding its limitations, this report has several strengths that enhance its teaching value. It documents a complete diagnostic arc — from an acute, alarming presentation, through a logically constructed differential and a small number of decisive investigations, to a specific aetiological diagnosis and a measurable clinical response — that mirrors the reasoning a clinician must perform in real time.[1,2] The concurrence of several classic elements in one patient (a movement disorder, cerebellar ataxia, a positive Chvostek sign, a suppressed PTH with profound hypocalcaemia and unmistakable bilateral calcification) makes the case an efficient illustration of the syndrome's core features, while the demonstrable reversibility provides a hopeful and clinically actionable message.[5,11] Presenting the biochemical, radiological and temporal data through clear tables and figures is intended to make the pattern immediately recognisable to readers who may encounter their first such patient without the benefit of prior experience.[10]

A further practical point concerns the differentiation of the movement disorder at the bedside. In a young woman taking multiple central-nervous-system-active medications, acute involuntary movements could plausibly be attributed to drug-induced dyskinesia, a functional movement disorder, an acute dystonic reaction, a metabolic movement disorder such as the hemichorea seen with deranged glucose metabolism, or a primary paroxysmal movement disorder, and each of these might have prompted a

very different management path.[1,5,25] The decisive clue in this patient was the accompanying neuromuscular irritability — a positive Chvostek sign — which reframed the presentation as a manifestation of hypocalcaemia and directed investigation towards calcium metabolism, whereupon the CT and biochemistry rapidly confirmed the diagnosis.[11,14] This illustrates a broader clinical principle: signs of latent tetany should always be sought in a patient with an unexplained movement disorder or seizures, especially against a background of thyroid or parathyroid surgery, because they may be the only immediate pointer to a treatable metabolic cause.[1,6]

The neuropsychiatric and quality-of-life dimensions of hypoparathyroidism deserve emphasis, because they are easily overlooked when attention is focused on the movement disorder. Chronic hypoparathyroidism is associated with fatigue, anxiety, depression and cognitive complaints that materially reduce quality of life, and validated disease-specific questionnaires are increasingly recommended for monitoring.[16,24] In Fahr syndrome specifically, cognitive impairment ranging from mild memory disturbance to dementia and a spectrum of psychiatric syndromes can dominate the picture, sometimes preceding the movement disorder by years.[11,17] Our patient's minor memory impairment, documented at presentation, is consistent with early cortical and subcortical involvement and provides a rationale for longitudinal neurocognitive follow-up even after her motor symptoms resolved.[2]

Although the surgical history made post-thyroidectomy hypoparathyroidism the obvious cause in this patient, a complete assessment of acquired hypoparathyroidism ordinarily considers autoimmune, infiltrative and genetic contributors as well, because these carry different implications for surveillance and family screening.[5,10] Autoimmune hypoparathyroidism, whether isolated or part of an autoimmune polyendocrine syndrome, is the commonest non-surgical cause in adults and may be accompanied by other endocrine deficiencies; infiltrative disorders and, rarely, mitochondrial disease such as MELAS can also impair parathyroid function and produce Fahr-type calcification.[2,13] In the present case, the clear temporal relationship between total thyroidectomy and the onset of hypocalcaemia, together with the absence of features suggesting a polyglandular or mitochondrial disorder, made the post-surgical mechanism secure; nonetheless, the breadth of this differential reinforces why a structured aetiological work-up, as outlined in Table 4, should accompany every new diagnosis of Fahr syndrome.[17,20]

This patient's course also carries a broader diagnostic and preventive message. Post-surgical hypoparathyroidism is the single commonest cause of chronic hypoparathyroidism and was the operative mechanism in this patient, developing after total thyroidectomy exactly as documented in other post-surgical Fahr syndrome reports.[8,10] Routine post-thyroidectomy surveillance of calcium and PTH, with timely calcium and active vitamin D supplementation, plausibly reduces the risk of chronic hypocalcaemia and its long-term sequelae, of which Fahr syndrome is among the most serious.[8,10] Had this patient's calcium been rigorously controlled from the outset, and had neuroimaging been performed when she first presented with seizures, the diagnosis might have been reached years earlier and the extensive calcification perhaps mitigated. The report thus argues for phosphocalcic screening and a low threshold for cranial CT in any thyroidectomised

patient who develops new neurological symptoms, and against reflexively labelling such seizures "idiopathic".[1,10]

From a health-systems perspective, this case carries lessons that extend beyond the individual patient, particularly for settings such as Indonesia where documented experience of Fahr syndrome remains limited and access to specialist neuroimaging and biochemical testing can be uneven.[10] The diagnosis in this patient ultimately required only two widely available investigations — a non-contrast head CT and a measurement of serum calcium and PTH — yet these had not been performed during the six years in which she was treated for presumed idiopathic epilepsy. Embedding a simple reflex into routine care, namely checking calcium metabolism and considering cranial imaging in any thyroidectomised patient who develops seizures, a movement disorder or cognitive change, would allow many such diagnoses to be made far earlier and at modest cost.[1,2] Equally, structured post-thyroidectomy follow-up that includes calcium and PTH surveillance would identify chronic hypoparathyroidism before its neurological complications accrue.[8,10] Reporting cases from under-represented regions helps to build the local evidence base needed to justify and design such pathways.[5,10]

The prominent cerebellar involvement in this patient is worth underscoring, both radiologically and clinically. While basal-ganglia calcification is the best-known feature of Fahr syndrome, deposition within the dentate nuclei and cerebellar hemispheres is well described and, when extensive, produces the ataxia and dysarthria that were evident here.[1,2] The coexistence of supratentorial and infratentorial calcification in a single patient signals a heavy overall calcification burden and correspondingly greater clinical severity, consistent with the reported relationship between calcification extent and phenotype.[5,7] The resolution of her ataxia with metabolic correction, in parallel with the improvement in her hyperkinetic movements, suggests that the cerebellar dysfunction was at least partly functional and reversible rather than reflecting fixed structural damage, an encouraging observation that aligns with the broader message of treatability in secondary Fahr syndrome.[1,2]

Comprehensive diagnosis and management of this patient therefore emphasise the importance of early detection and appropriate treatment of Fahr syndrome, particularly when it is related to disorders of calcium metabolism.[1] This report contributes novelty by identifying a rarely documented finding, especially in the Indonesian population: a bilaterally symmetrical calcification distribution involving not only the basal ganglia but also the parietal cortex and both cerebellar hemispheres in a patient with post-thyroidectomy hypoparathyroidism. At the same time, the report has limitations. Data on cognitive function and several specific laboratory values — notably serum phosphate, magnesium and vitamin D — were not available, so the biochemical–metabolic correlation could not be analysed in depth, and it was not recorded whether the reported serum calcium was albumin-corrected or ionised. The chronic diagnosis of hypoparathyroidism rested on the longitudinal clinical course rather than on repeated contemporaneous biochemistry, and the historical seizure semiology was not detailed in the available records; the reinterpretation of the earlier epilepsy as hypocalcaemic in origin, though highly plausible, is therefore necessarily retrospective and cannot be proven. Electroencephalography, magnetic resonance imaging, dopamine-

transporter imaging and repeat CT were not performed, so the electroclinical and radiological course could not be corroborated with these modalities. In addition, long-term monitoring of the progression of calcification could not be fully evaluated within the available follow-up period, and the response to therapy was assessed clinically rather than with repeat imaging or formal neurocognitive testing. The acute deterioration is best understood as an exacerbation of chronic hypocalcaemia, plausibly precipitated by fatigue as the history suggests, rather than as a new and separate process.

Taken together, this case exemplifies how a single, correctable metabolic lesion can present through several apparently unrelated neurological syndromes — a seizure disorder, a hyperkinetic movement disorder, cerebellar ataxia and mild cognitive impairment — that are unified by chronic hypoparathyroid hypocalcaemia and its radiological footprint of widespread intracerebral calcification. The favourable response to conventional metabolic therapy and anticonvulsant optimisation supports the central thesis that timely recognition converts a potentially disabling neurodegenerative-appearing illness into a treatable one, while the extent of calcification already present serves as a cautionary marker of the years of uncontrolled hypocalcaemia that preceded diagnosis.[1,2,7]

4. Conclusion

Fahr syndrome is a rare disorder characterised by bilateral, symmetrical calcification of the basal ganglia, cerebellum and cerebral cortex, giving rise to movement disorders, seizures and cognitive impairment. We described a 35-year-old woman who, seven years after total thyroidectomy, presented with involuntary movements, tremor, dystonia, ataxia and a positive Chvostek sign; non-contrast CT revealed multiple bilateral calcifications, while laboratory testing confirmed severe hypocalcaemia and low PTH, establishing a diagnosis of secondary Fahr syndrome due to post-thyroidectomy hypoparathyroidism. Management with calcium supplementation, active vitamin D, the anticonvulsant phenytoin and supportive therapy, together with attention to motor rehabilitation, produced near-complete clinical recovery within six weeks that was sustained at two months. Although the established calcification itself cannot currently be reversed, prompt and early intervention can arrest further progression and substantially improve the patient's neurological function and overall quality of life.

Accordingly, routine measurement of serum calcium, phosphate and PTH is recommended in patients who have undergone thyroidectomy, and neuroimaging should be considered before a seizure disorder is labelled idiopathic in such patients. The case also demonstrates that a diligent aetiological work-up — distinguishing secondary Fahr syndrome from primary familial brain calcification and from the genetic, infectious and toxic mimics — is essential, because it defines both the treatment target and the need for family screening. Above all, it shows that sustained metabolic control functions simultaneously as treatment for existing symptoms and as secondary prevention against further calcification and neurological deterioration. Future case reports should ideally include longitudinal data on neurocognitive evaluation, serial imaging and treatment response, and further research is needed to clarify the risk factors, the molecular mechanisms of calcification, and the comparative effectiveness of the various metabolic interventions

— including PTH-replacement therapy — available for the hypoparathyroid population.

Declarations

Ethics approval and consent to participate

The study was conducted in accordance with the principles of the Declaration of Helsinki. Reporting of this single case was approved by the institutional review process of Klungkung Regional General Hospital.

Consent for publication

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. Every effort has been made to protect the patient's identity; all direct identifiers have been removed from the imaging.

Availability of data and materials

The clinical data supporting the findings of this case report are included within the article. Further detail is available from the corresponding author on reasonable request, within the constraints of patient confidentiality.

Competing interests

The authors declare that they have no competing interests.

Funding

This case report received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

Authors' contributions

Both authors were involved in the clinical care of the patient, the acquisition and interpretation of the clinical data, and the drafting and critical revision of the manuscript. Both authors read and approved the final version.

Acknowledgements

The authors thank the patient for her kind consent to share this instructive case.

5. References

1. Berrabeh S, Messaoudi N, Elmehraoui O, et al. Hypoparathyroidism and Fahr's syndrome: a case series. Cureus. 2023;15(6):e40502. doi:10.7759/cureus.40502
2. Srichawla BS, Andrade E, Kipkorir V. Decoding brain calcifications: a single-center descriptive case series and examination of pathophysiological mechanisms. SAGE Open Med Case Rep. 2023;11:2050313X231197521. doi:10.1177/2050313X231197521
3. Tekin Orgun L, Besen Ş, Sangün Ö, et al. First pediatric case with primary familial brain calcification due to a novel variant on the MYORG gene and review of the literature. Brain Dev. 2021;43(7):789–797. doi:10.1016/j.braindev.2021.04.002
4. Al Ali J, Yang J, Phillips MS, et al. A case report of a patient with primary familial brain calcification with a genetic variant. Front Neurol. 2023;14:1235909. doi:10.3389/fneur.2023.1235909
5. Kalampokini S, Georgouli D, Dadouli K, et al. Fahr's syndrome due to hypoparathyroidism revisited: a case of parkinsonism and a review of all published cases. Clin Neurol Neurosurg. 2021;202:106514. doi:10.1016/j.clineuro.2021.106514
6. Arruda ACG de, Guerra ACDZ, Pessoa CH, et al. Hypoparathyroidism and Fahr's syndrome: case series. J Bras Nefrol. 2022;44(4):592–596. doi:10.1590/2175-8239-JBN-2020-0243
7. Zavatta G, Tebben PJ, McCollough CH, et al. Basal ganglia calcification is associated with

local and systemic metabolic mechanisms in adult hypoparathyroidism. J Clin Endocrinol Metab. 2021;106(7):1900–1917. doi:10.1210/clinem/dgab162

8. Alrashidi FS, Mohajab AH. Basal ganglia calcification secondary to postsurgical hypoparathyroidism presenting with seizures and acute confusion: a case report (Fahr syndrome). Cureus. 2026;18(1):e100553. doi:10.7759/cureus.100553
9. Kar P, Goswami R. Effect of calcitriol and calcium on basal ganglia calcification in hypoparathyroidism: experimental models. J Mol Endocrinol. 2022;70(2):e220108. doi:10.1530/JME-22-0108
10. Supit VD, Kurniawan D, Fatimah E. Fahr syndrome and neurological manifestations in hypoparathyroidism patients. Radiol Case Rep. 2024;19(4):1248–1253. doi:10.1016/j.radcr.2023.12.034
11. Mohammedin AS, Alkharashi AF, Alabdulqader AA, et al. Fahr's syndrome presenting with hypocalcemia and psychotic features. Cureus. 2021;13(9):e18091. doi:10.7759/cureus.18091
12. Madadi S, Alami K, Kaneda Y, et al. From seizures to cognitive dysfunction: a case report of Fahr syndrome in an Afghan patient. Medicine (Baltimore). 2024;103(25):e38542. doi:10.1097/MD.0000000000038542
13. Li J, Wang X, Guo Y, et al. Fahr syndrome, hypoparathyroidism and mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes (MELAS) syndrome. AACE Endocrinol Diabetes. 2025;13(1):102–106. doi:10.1016/j.aed.2025.09.007
14. Le Hir A, Hak JF, Gragueb-Chatti I, et al. Hypocalcemia-induced seizure with Fahr's syndrome. J Nephrol. 2022;35(3):1047–1048. doi:10.1007/s40620-022-01260-w
15. Bitew HY, Kambutse I, Tuyizere A, et al. Fahr syndrome presenting with status epilepticus after COVID-19 infection. JCEM Case Rep. 2023;1(3):luad072. doi:10.1210/jcemcr/luad072
16. Manna R, Roy A, Das A, et al. Cerebral calcification and treatment-resistant seizures: a rare syndromic presentation of pseudohypoparathyroidism — a case report. AACE Endocrinol Diabetes. 2025;13(1):98–101. doi:10.1016/j.aed.2025.09.016
17. Zalewska E, Gnacińska-Szymańska ME, Obołończyk Ł, et al. Coexistence of DiGeorge syndrome with Fahr syndrome, mosaic Turner syndrome and psychiatric symptoms — a case report. Psychiatr Pol. 2021;55(2):397–404. doi:10.12740/PP/119376
18. Wang X, Xu T, Zhu Y, et al. Fahr's syndrome with pseudohypoparathyroidism: oral features and genetic insights. Int J Mol Sci. 2024;25(21):11611. doi:10.3390/ijms252111611
19. Zhang M, Jia Y, Wang H, et al. Epileptic seizure as the prominent symptom in Fahr syndrome, case report and literature review. Seizure. 2024;123:88–91. doi:10.1016/j.seizure.2024.09.023
20. Palu G, Moraes ST, Romaniello G, et al. Could Fahr's syndrome have more than one simultaneous etiology? Cureus. 2021;13(12):e20342. doi:10.7759/cureus.20342
21. Jithesh G, Bansal R, Ajmal M, et al. Young male with seizure disorder and intracranial calcification: a case of Fahr's syndrome. Cureus. 2022;14(2):e22189. doi:10.7759/cureus.22189
22. Sarna MK, Goel P, Bhargava V, et al. Fahr's syndrome associated with

hypoparathyroidism: a case report. J R Coll Physicians Edinb. 2023;53(4):283–287. doi:10.1177/14782715231210606

23. Iwase T, Yoshida M, Iwasaki Y, et al. Selective extension of cerebral vascular calcification in an autopsy case of Fahr's syndrome associated with asymptomatic hypoparathyroidism. Neuropathology. 2021;41(5):387–395. doi:10.1111/neup.12760
24. Mohammed YE, Gebreselassie KH, Sesega EH, et al. Recurrent seizures in an adolescent female with extensive intracranial calcifications: a case report of Fahr's syndrome secondary to hypoparathyroidism. Cureus. 2025;17(2):e78780. doi:10.7759/cureus.78780
25. Oksenhendler R, Pellerin D, Almutlaq A. A case of non-ketotic hyperglycemic hemichorea and Fahr syndrome. Cureus. 2024;16(5):e60265. doi:10.7759/cureus.60265