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Bilateral Knee Synovial Chondromatosis in a 2-Year-Old Girl: First Reported Case and Literature Review

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 2-year-old
Final Diagnosis: Bilateral knee synovial chondromatosis
Symptoms: Joint effusion • limited range of motion • limping gait • pain in the knee joint
Clinical Procedure: —
Specialty: Orthopedics and Traumatology

Objective: Rare disease

Background: Synovial chondromatosis (SC) is a rare monoarticular benign condition characterized by the development of intra- or periarticular cartilage-like nodules or loose bodies. Typically affecting middle-aged men, SC is exceptionally rare in female pediatric patients. Symptoms include diffuse pain, swelling, and limited joint movement. The loose bodies are typically numerous, small, rounded, and uniform in size. Histologically, they consist of hyaline cartilage-like nodules containing clusters of chondrocytes. Bilateral SC in the knee of young children is even rarer and has not been previously documented in the medical literature.

Case Report: A 2-year-old girl presented with recent pain in her right knee without preceding trauma. Physical examination revealed joint effusion, a limping gait, and limited knee flexion, with no signs of local inflammation. Radiographs showed normal joint structures, and laboratory test results, except for an elevated CRP, were normal. Joint aspiration ruled out septic arthritis. The worsening of symptoms within a relatively short period prompted further investigation, leading to the identification of numerous cartilaginous nodules within the joint during exploratory arthrotomy. Microscopic examination revealed unossified cartilage-like nodules containing disorganized chondrocytes. Four months after surgery, she was diagnosed with SC in the contralateral knee.

Conclusions: Given the rarity of synovial chondromatosis in young children, and the even rarer bilateral presentation in a large joint, this case report provides an opportunity to explore the challenges in diagnosing and managing such conditions in pediatric patients. To the best of our knowledge, this is the youngest reported case of bilateral SC of the knee in a pediatric patient.

Keywords: Knee Joint • Chondromatosis, Synovial • Joint Loose Bodies

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Introduction

Synovial chondromatosis (SC), also known as synovial osteochondromatosis, is a rare benign condition characterized by the development of cartilage-like nodules or loose bodies within the synovial lining of joints, bursae, and tendon sheaths [1]. The disease typically affects a single large joint, most commonly targeting the knee in adults between their third and fifth decades of life [2]. Bilateral involvement, particularly of the knee joints, is even rarer [3], and is usually documented in adult patients with other underlying inflammatory articular conditions [4,5]. SC occurs more often in men, is seldom observed in individuals under the age of 20, and monoarticular and bilateral presentations are particularly uncommon in children, especially girls, in their early years of life [6,7].

The initial clinical presentation of unilateral or bilateral synovial chondromatosis (SC) is often nonspecific. While some patients are asymptomatic, those with symptoms typically report diffuse pain worsening with movement, local swelling, decreased range of motion, and joint blockage [2,8,9]. A key radiographic finding is multiple rounded intra- or periarticular cartilaginous nodules. However, as calcification occurs in only 70% of cases and is time-dependent, relying solely on radiographs may yield inconclusive results [10]. In such cases, computed tomography (CT) or magnetic resonance imaging (MRI) can provide a clearer view of the joint surfaces and surrounding structures, allowing for a more accurate assessment of the disease extent [1,11].

Synovial chondromatosis can manifest as a primary form, with no evidence of concurrent joint pathology, or as a secondary form resulting from pre-existing joint conditions. In both cases, the condition is characterized by the formation of multiple chondroid nodules [8,9]. The cartilaginous loose bodies originate as sessile nodules in synovial membranes, bursae, and tendon sheaths as a result of metaplasia [12]. As these nodules grow, they detach and become free intra- or periarticular bodies. Typically, the loose bodies are numerous, small, rounded, and uniform in size, with a smooth, pearly white surface. Histologically, they are characterized by hyaline or myxoid cartilage-like nodules containing clusters of chondrocytes, which may eventually exhibit endochondral ossification [12,13].

Although SC can sometimes be managed conservatively, surgical removal of the intra-articular cartilaginous bodies is typically the definitive treatment. The preferred approach involves arthroscopic or open removal of loose bodies, with or without synovectomy. While synovial membrane removal can reduce the likelihood of disease recurrence, it could also increase the risk of joint and periarticular damage over time [14]. The prognosis varies depending on the severity of the disease, but in general, prompt diagnosis followed by early intervention can prevent the progression to a more debilitating form of the disease [3,15,16]. Recurrence is possible, mostly as the result of incomplete removal of the loose bodies; therefore, long-term follow-up is advised to monitor for any new development of loose bodies or joint damage [17].

The occurrence of SC in children, particularly in infants and toddlers, is exceedingly rare. Although there are reported cases in children under 10 years old [6,18-24], the incidence of bilateral disease in early childhood has not been previously documented. In this report, we present the case of a 2-year-old girl experiencing debilitating pain in her right knee, ultimately diagnosed with SC during an exploratory arthrotomy. Four months after surgery on the right knee, she was diagnosed with SC in the contralateral knee.

Case Report

A 2-year-old girl presented with a recent history of pain in the right knee joint, without any significant preceding trauma or fever. Despite prior treatment with dexamethasone, she did not experience any improvement in her symptoms. On physical examination, the patient exhibited joint effusion, a limping gait, and a limited range of motion, particularly in flexion, of the right knee. There were no signs of local inflammation, and her family history was negative for rheumatological diseases. Plain radiographs of the right knee showed normal joint structures without any signs of intra-articular or bony lesions. Laboratory tests, including markers for autoimmune diseases, returned normal results. Due to elevated C-reactive protein (CRP) levels, a joint aspiration was performed to rule out septic arthritis. Synovial fluid analysis excluded the diagnosis of infectious arthritis (**Table 1**). The child was discharged, and her mother was advised to return for a clinical review in 2 weeks. Approximately 10 days after the initial consultation, the patient returned with tenderness and mechanical locking of the right knee. Subsequently, an exploratory arthrotomy was performed through a lateral suprapatellar approach for further investigation. During the surgery, numerous small cartilaginous nodules were identified inside the right knee joint, displaying an elastic consistency and smooth, pearly white surface (**Figure 1**). Microscopic examination of the retrieved loose bodies revealed well-demarcated, unossified nodules with disorganized clusters of small chondrocytes within a basophilic chondroid matrix in the central regions. More peripherally, the nodules were surrounded by deposits of fibrin-like material (**Figure 2**). Postoperative recovery proceeded uneventfully, and the patient was discharged from the hospital after a short stay and remained asymptomatic. Four months after the surgical procedure on the right knee, the child returned with pain and moderate effusion in the left knee, associated with synovitis (**Figure 3**). She was treated with a 3-day course of oral prednisone, which did not improve the joint effusion and

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Table 1. Initial blood test.

Compound	Result	Reference range
C-reactive protein (mg/L)	23.1	<5
Anti-CCP antibodies (U/mL)	5.89	<20
Anti-Ro/SSA antibodies (U/mL)	<0.4	<7
Anti-La/SSB antibodies (U/mL)	<0.4	<7
Rheumatoid factor (U/mL)	<8	<20
Synovial fluid culture	Negative for bacteria or fungi growth	–

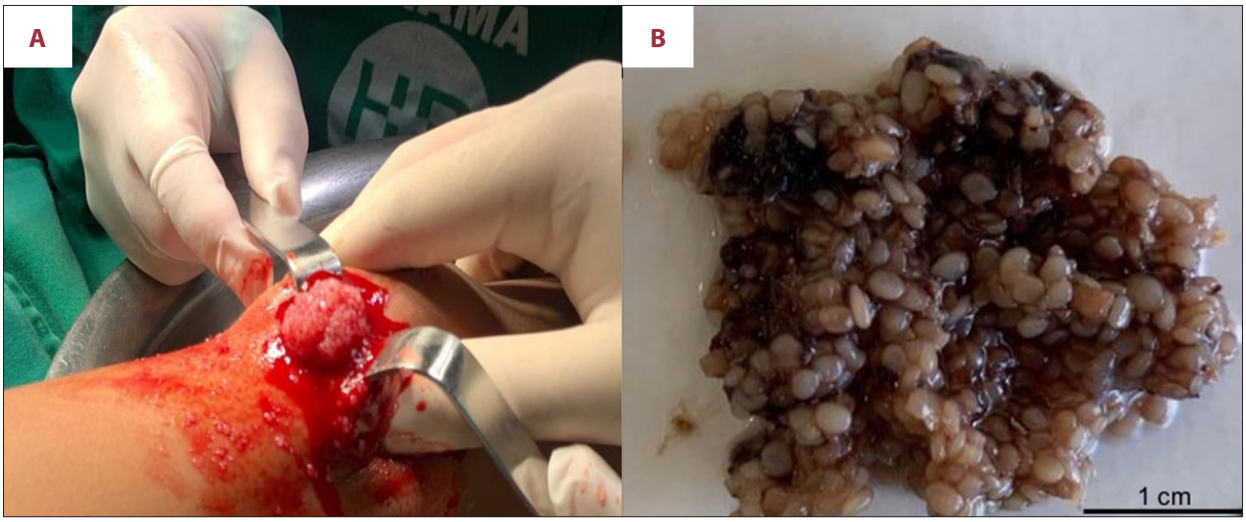

Figure 1. Cartilaginous loose bodies in primary synovial chondromatosis. (**A**) The intraoperative image illustrates numerous tiny cartilaginous nodules removed from the right knee joint space. (**B**) Gross examination reveals that the formalin-fixed nodules have smooth surfaces and exhibit elastic consistency.

pain. A lateral suprapatellar arthrotomy was subsequently performed on the left knee, revealing numerous cartilaginous loose bodies in the joint space. Macroscopic and histological findings were similar to the cartilaginous nodules previously identified in the right knee (**Figure 4**). Five weeks after the second surgery, the child was free from any articular problems.

Discussion

Biarticular involvement in SC is extremely rare. To our knowledge, only 6 cases of bilateral knee involvement have been documented in the literature. These include a 14-year-old boy [25], an 18-year-old male with genu valgum deformity [5], a 34-year-old woman with rheumatoid arthritis [4], and 3 previously healthy individuals aged 30 [3], 38 [26], and 67 years [27]. Consequently, our case of bilateral presentation in a 2-year-old girl is exceptionally rare.

Bilateral SC shares similar clinical and demographic characteristics with unilateral SC. Although its pathogenesis remains debatable, SC is generally considered a benign metaplastic disease [28-30]. However, its neoplastic nature has been suggested based on observed clonal chromosomal changes in some cases [31] and rare reports of malignant transformation [32], especially in highly cellular lesions or cases associated with recurrence or worsening symptoms [29,30].

In this case report, the patient's initial symptoms of knee joint pain, along with joint effusion, limping gait, and limited range of motion without a history of trauma, posed a diagnostic challenge. Initial presentations of pain, swelling, and restricted range of motion could easily be attributed to more common pediatric knee disorders, leading to delays in the correct diagnosis. This misdiagnosis could result in inappropriate treatment strategies, rather than more definitive surgical interventions. This clinical presentation is nonspecific and consistent with several proliferative conditions of the synovial membrane. Therefore, the differential diagnosis can include chronic synovial proliferation, pigmented villonodular synovitis, infectious arthritis, juvenile idiopathic arthritis, or even malignancies. Clinical and diagnostic similarities with these

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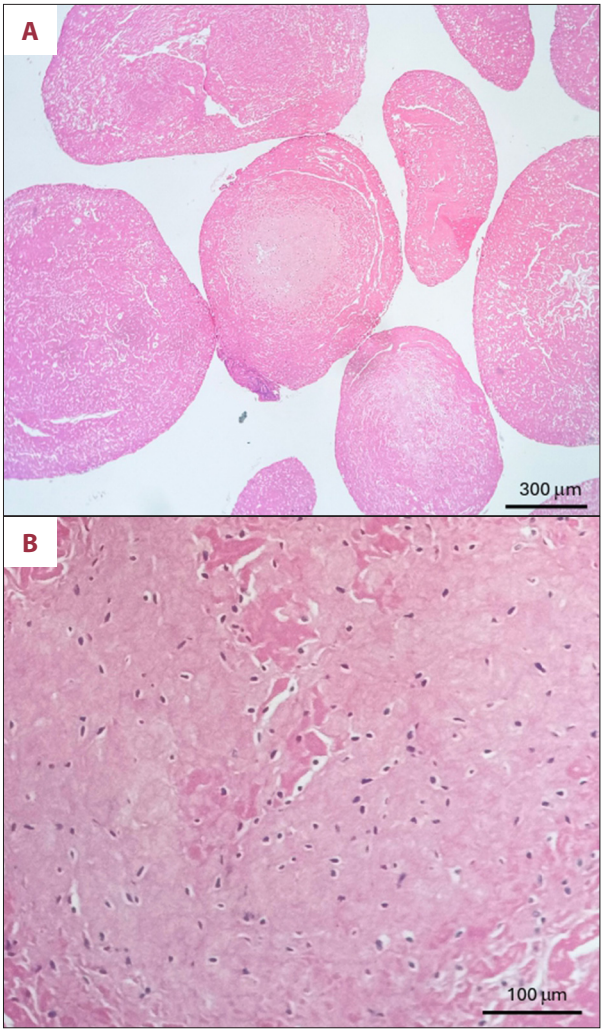

Figure 2. Microscopic examination of the retrieved loose bodies from the right knee. (**A**) Well-demarcated unossified nodules. (**B**) Disorganized clusters of small chondrocytes located in the central regions of the nodules. (**C**) Chondrocytes embedded within a non-calcified basophilic matrix. H&E staining.

articular conditions are notable, especially when SC presents without calcified bodies, as seen in this case [33,34]. In particular, the differential diagnosis between SC and juvenile idiopathic arthritis can be challenging for clinicians, as both conditions can present with similar symptoms [35]. Misdiagnosis can lead to inappropriate or unnecessary treatments, resulting in overtreatment and potential harm, particularly in pediatric patients. Therefore, a careful clinical evaluation – encompassing a comprehensive patient history, laboratory tests, and imaging – is crucial for making an accurate diagnosis and minimizing the risks of mismanagement.

Despite extensive clinical and laboratorial evaluation, including synovial fluid analysis and imaging studies, the initial diagnosis in the right knee remained elusive. In our patient, the lack of calcification in the nodules prevented their detection on plain radiographs. Indeed, SC may go unnoticed on initial radiographs, as 5-30% of cases do not exhibit nodular calcification [10]. Although MRI has been a useful diagnostic tool for evaluating soft tissue involvement when calcification is absent [34], it may not always provide definitive evidence of SC, especially in early stages when the disease can present with subtle changes. The potential for imaging findings to overlap with other pediatric knee pathologies complicates diagnosis, especially in young children who may have less typical clinical presentations. A case of SC was reported in a 10-year-old girl whose MRI findings did not confirm the diagnosis of SC [21]. In this case, the images failed to distinguish SC from other proliferative conditions of the synovium. Similar to our report, the definitive diagnosis was established based on gross and histological aspects.

According to Milgram [36], the process of nodule formation in SC consists of 3 phases: the initial stage is characterized by active disease solely affecting the synovial membrane, transitional lesions exhibiting intrasynovial activity as well as loose free bodies, and a third phase wherein numerous free bodies are identified in the joint. Macroscopically, the cartilaginous free bodies can range from tiny nodules as small as 1 mm to larger nodules exceeding 1 cm, found anchored to the synovium or freely suspended within the synovial fluid [18]. In our case, the identification of numerous small cartilaginous nodules inside both knee joints during exploratory arthrotomies confirmed the diagnosis of sequentially bilateral SC. In both knees, the nodules were abundant and exceedingly tiny and were easily removed from the intra-articular space during the surgical procedure. Microscopic examination of the retrieved loose bodies from both knees revealed characteristic features consistent with SC, including well-demarcated unossified nodules with clusters of chondrocytes surrounded by a basophilic chondroid matrix.

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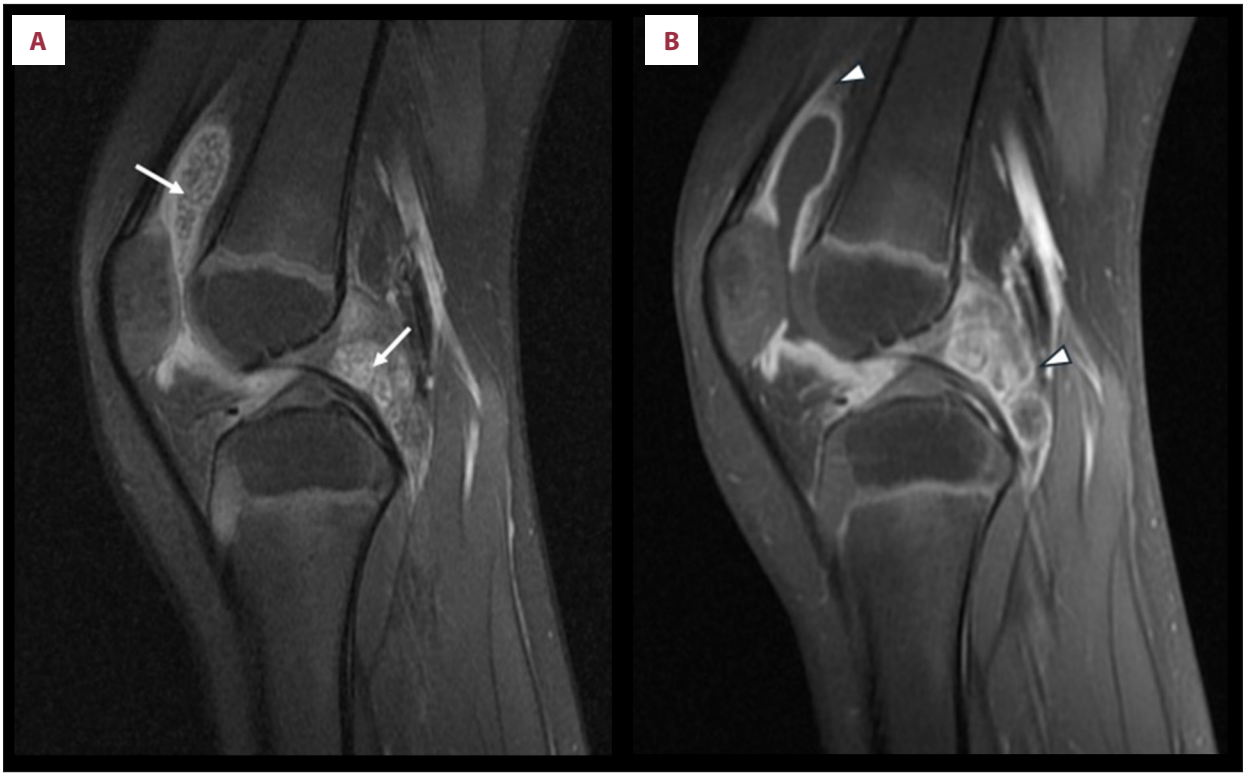

Figure 3. MRI scans with proton density fat suppression (**A**) and T1-weighted imaging with fat suppression after contrast (**B**) in the sagittal plane showing multiple foci of hyaline cartilage (arrow) associated with synovial thickening and contrast enhancement (arrowhead) in the suprapatellar recess and the posterior joint recess of the knee.

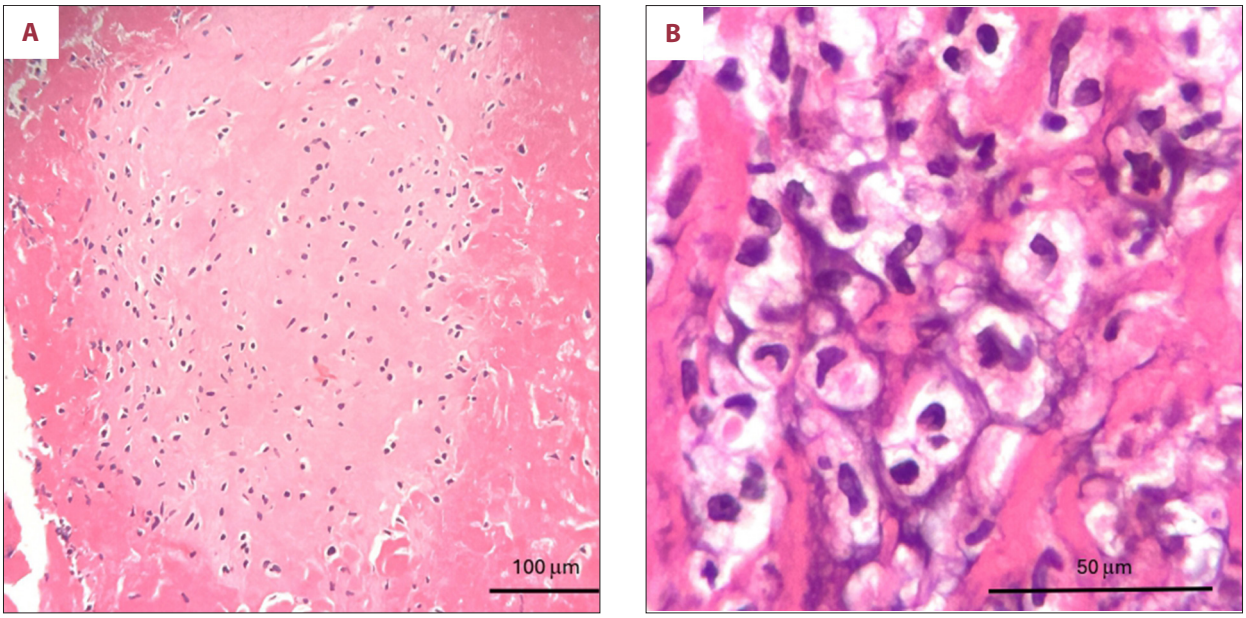

Figure 4. Microscopic examination of the retrieved loose bodies from the left knee. (**A**) Clusters of small chondrocytes, irregularly arranged, are present in the central area of the nodule. (**B**) Hypercellular cartilage showing nuclear crowding and early signs of endochondral ossification. H&E staining.

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While most SC cases are benign, the potential for malignant transformation, although rare, should not be overlooked, particularly given the aggressive nature and poor prognosis that can accompany malignant variants such as chondrosarcoma. The reported relative risk of 5% is associated clinically with multiple recurrences and rapid worsening of symptoms [29,30]. Histological criteria for diagnosing malignant transformation include atypical chondrocytes dispersed in sheets throughout the ground substance [37], mitotic figures, myxoid change, necrosis, and bone permeation [29,30]. Wide en-bloc excision is the preferred surgical treatment for most secondary malignant lesions. Adjuvant therapies, such as chemotherapy and radiation, may be considered, although their effectiveness is variable and often debated in the literature [38]. In our case, despite the complexity of the initial presentation and diagnostic challenges, the patient's postoperative recovery from both surgeries was favorable. However, it should be noted that bilateral joint involvement can indicate a more severe form of SC that requires more precise and invasive treatment [3], posing additional therapeutic challenges in pediatric patients. Given the rarity of the condition in young children, there is a need for careful monitoring following surgical removal of loose bodies to prevent recurrence, joint degeneration, or functional impairment. Furthermore, long-term follow-up to assess the development of any late complications such as secondary osteoarthritis and regular imaging and clinical assessments are essential to track recovery and manage any potential recurrence or joint deterioration.

In summary, the findings from this case report emphasize the crucial importance of a collaboration between pediatric orthopedic surgeons, radiologists, rheumatologists, and physical therapists for accurate diagnosis, surgical planning, and postoperative care. A multidisciplinary team can help differentiate bilateral knee SC from other potential knee pathologies, optimize surgical outcomes, and provide holistic care for the child during both the acute and follow-up phases. This team-based approach ensures that all aspects of the child's health, including joint function, growth, and quality of life, are carefully considered throughout the treatment process. Further research and reporting of similar cases are warranted to enhance the understanding and management of SC in children.

Conclusions

Given the extreme rarity of bilateral synovial chondromatosis in young children, this case report provides an opportunity to explore the challenges in diagnosing and managing such conditions in pediatric patients.

Department and Institution Where Work Was Done

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Informed Consent

The study protocol was approved by the Centro de Estudos Superiores de Maceió review board and the research was conducted in accordance with the ethical principles outlined in the Declaration of Helsinki. Prior to participating, the child's mother provided written consent, by which she agreed to the use and analysis of her child's data.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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