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# Ormond's Disease – 26 Years of Experience at One Centre

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**Key words:** Ormond's disease – Periaortitis – Hydronephrosis – Immunosuppression

**Abstract:** Ormond's disease is a systemic autoimmune disease with serious complications. We present our retrospective analysis of 83 patients diagnosed with and treated for idiopathic retroperitoneal fibrosis (Ormond's disease) in our department from 1997 to 2023. In this retrospective study, we analysed the diagnostic approaches, the clinical history and surgical and immunosuppressive therapies, and their subsequent effects on our patients. Patients with established disease activity were given immunosuppressive treatment, using corticosteroids alone or in combination with azathioprine, in patients with exacerbation of the disease mycophenolate mofetil. Three patients with Ormond's disease and systemic complications (IgG4-related disease) were treated with rituximab. In the entire cohort, 83 patients received immunosuppressive therapy; the next 5 patients did not receive this treatment because they did not present inflammatory activity from the disease. In these 83 patients, computed tomography showed that immunosuppressive treatment resulted in partial or complete regression of the inflammatory infiltrate. Out of the 83 patients, 10 patients experienced disease exacerbation 7 and 24 months after the immunosuppressive treatment was discontinued. The follow-up ranged from 24 months to 26 years.

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## Introduction

Ormond's disease – idiopathic retroperitoneal fibrosis (IRF) – is a relatively rare disease with an unclear aetiology, characterised by chronic periaortitis and retroperitoneal fibrosis (RF). The inflammatory process affects the infrarenal part of the abdominal aorta and the iliac arteries. Moreover, infiltrates encasing the ureters and the inferior vena cava are also commonly found. Its incidence is 1.3 in 100,000 people (van Bommel et al., 2009). A subset of patients with Ormond's disease has immunoglobulin G4-related disease (IgG4-RD). The course of the disease is associated with the incidence of complications, the most serious of which are renal failure and aneurysm of the abdominal aorta or the iliac arteries. Thanks to further advances in medicine and diagnostics, therapeutic strategies are gradually changing. Improved results from pharmacotherapy regimens lead to a greater emphasis on early comprehensive medical therapy with corticosteroids and immunosuppressive or immunomodulatory drugs. In this retrospective study, we describe the clinical outcomes of 83 patients diagnosed with and treated for Ormond's disease in our department.

## Patients and Methods

### Patients and setting

Between 1997 and 2023, our department, in collaboration with the Department of Urology and the Department of Vascular Surgery, investigated 135 patients suspected to have Ormond's disease. Ultimately, 83 patients were diagnosed with and subsequently treated for this disease. Patients with RF that had been medically induced, caused by cancer, or exacerbated by other disease were excluded from the analysis. Ormond's disease was diagnosed based on a comprehensive clinical examination using biochemical, immunological, and microbiological tests, and imaging methods – ultrasound, computed tomography (CT), positron emission tomography (PET)/CT, and CT/arteriography. Laboratory screening included a biochemistry panel, screening for hepatitis B and C; the serum levels of IgG, specifically the IgG1-IgG4 subclasses; and the erythrocyte sedimentation rate (ESR). Patients were also examined for the presence of antinuclear antibodies, extractable nuclear antibodies, and antibodies to double-stranded DNA. RF was considered if a soft-tissue density surrounded the infrarenal aorta or iliac vessels on contrast-enhanced CT and/or on histological confirmation. Disease activity was confirmed by a PET/CT scan in all patients upon the initiation and discontinuation of immunosuppressive therapy. In 45 patients (54%), a biopsy was performed percutaneously using a biopsy needle under CT navigation, whereas in 3 patients, laparoscopy was used to collect the sample. The diagnosis of IgG4-RD in 28 patients was confirmed by subsequent histological examination: the presence of at least one characteristic histopathological feature, >30 $IgG4^+$ plasma cells per high power field, and an $IgG4^+/IgG^+$ plasma cell ratio cut-off of >40%.

### Treatment

All medical and surgical treatments for IRF were reviewed. The medications used included glucocorticoids, azathioprine, mycophenolate mofetil, and rituximab. The first-line treatment was a combination of corticosteroids and azathioprine or corticosteroids alone. Mycophenolate mofetil therapy was subsequently used in 5 of these patients with exacerbations of the disease. Rituximab was used in three patients with systemic clinical manifestations associated with IgG4-RD and exacerbations of IRF after finishing of the corticosteroids therapy. Where indicated, surgical procedures were applied, namely ureteral stenting, nephrostomy, stenting of arteries.

### Follow-up and outcomes

Patients in the study were followed-up regularly during the treatment of their disease (via medical record review) and then at least once per year for the next 5 years. The primary outcome was the reduction of clinical symptoms, extubation of the obstructed ureters, and reduction of soft tissue mass, followed by the absence of inflammatory activity on a PET/CT scan. The secondary end-points included monitoring the biochemical parameters of renal function, the IgG4 concentration, and reaching normal values of inflammatory biomarkers.

The patients were monitored at regular intervals of 2–4 weeks during the first 4 months when biochemical and immunological parameters were monitored. After inflammatory markers became insignificant, this interval was extended to 2 months. Follow-up CT or PET/CT scans were performed after the first 6 months and then as needed. The follow-up range was 24 months to 26 years.

## Results

We investigated 135 patients between July 1997 and June 2023 who were suspected to have IRF. We excluded 52 patients: 5 patients without activity of the disease, 34 patients did not have Ormond's disease, 8 patients had an inflammatory abdominal

aortic aneurysm, and 5 patients had a cancerous retroperitoneal mass. Our final study population included 83 patients, 55 (66%) of whom were men and 28 (24%) of whom were women. All patients had imaging findings consistent with Ormond's disease. The diagnosis was confirmed by biopsy in 48 cases (58%). Fifty-three patients presented with clinical symptoms of ureteric obstruction with hydronephrosis, renal insufficiency (17 patients), or renal failure (5 patients). Thirty patients had periaortitis, with aneurysms of the aorta and/or iliac arteries present in 12 of these patients. After laboratory testing, 22 patients were positive for antinuclear antibodies, without evidence of specificity for extractable nuclear antibodies. None of the patients tested positive for antibodies to double-stranded DNA. Other autoimmune diseases were present in 25 patients, in which 14 had autoimmune thyroiditis, 2 had vasculitis, and 9 had Sjogren's syndrome. The IgG4 concentration was examined in 52 patients; 28 had an elevated IgG4 serum concentration. The IgG4 serum concentration was within the normal range in all patients after receiving immunosuppressive therapy. Hydronephrosis was found in 32 patients; 18 patients showed impairment of one ureter and 14 patients had impairment of both ureters. In 7 patients, despite an initial diagnosis of renal insufficiency, subsequent surgical intervention and immunosuppressive therapy resulted in the recovery of renal function. The standard surgical solution involved the insertion of stents, and a nephrostomy was performed on 3 patients. In all 83 patients, CT showed that immunosuppressive treatment resulted in partial or complete regression of the inflammatory infiltrate. After termination of the immunosuppressive therapy, 32 ureters were extubated successfully. Eight patients presented an abdominal aortic aneurysm, and 6 patients had iliac artery aneurysms. Surgical treatment was indicated for aneurysms in 4 patients prior to the deployment of immunosuppressive therapy. In 1 patient with an aneurysm, surgical treatment was not indicated after the initial diagnosis, but 4 months after termination of immunosuppressive treatment, the size of the aneurysm had increased, and surgery was required. Four patients who underwent surgery for aneurysms were subsequently given standard immunosuppressive treatment, with no exacerbation of the disease for 6 months after its termination.

83 patients received immunosuppressive therapy; the next 5 patients did not show inflammatory activity from the disease, so they did not receive this treatment. The 83 treated patients received a combination of corticosteroids with azathioprine, corticosteroids or mycophenolate mofetil alone. Out of the 83 patients, 10 patients (12%) experienced disease exacerbation 7 and 24 months, respectively, after the immunosuppressive treatment was discontinued. There was no difference in the success of therapy between the IgG4-associated and non-IgG4-associated groups of patients with Ormond's disease.

The follow-up period ranged from 24 months to 26 years. Out of the total number of 83 patients, 80 are still alive. One patient died of an acute abdominal event, and 2 patients died of cancer.

## Discussion

### Disease diagnosis

IRF has an unclear aetiology. The term covers several common diseases – chronic periaortitis, inflammatory abdominal aortic aneurysm, and perianeurysmal RF. There is a general consensus that IRF is an autoimmune disease. The reported autoantigen is an atherosclerotic plaque (Parums, 1990). Some patients with Ormond's disease meet the definition of IgG4-RD (Khosroshahi et al., 2013). One of the complex and very important issues to be addressed in the diagnosis of RF involves distinguishing between the infectious and non-infectious aetiology of the inflammation. The emergence of a retroperitoneal or para-aortal infiltrate may also be induced by an infectious aetiology. Given the use of immunosuppressive therapy, the failure to recognise this difference may have fatal consequences for the patient. Potential causative agents may include viruses (e.g., hepatitis), mycobacteria, *Staphylococcus aureus*, and *Salmonella* (Sekar, 2010; Cartery et al., 2011). In our patient cohort, we did not observe differences in clinical symptomatology in patients meeting the histological criteria of IgG4-RD compared with patients without evidence of IgG4-RD. In most patients with IgG4-RD, the IgG4 serum concentration did not correlate with positive histological findings based on biopsy. This agrees with our findings in patients with abdominal aortic aneurysm (Hao et al., 2016; Prucha et al., 2019). We tried to establish a histological diagnosis for all patients by performing CT guided biopsy or, in three cases, laparoscopically. In other cases, obtaining a sample was associated with excessive risk to the patients due to the location or time constraints. In these instances, the laboratory results and imaging findings were correlated to confirm the diagnosis. The other possibility of diagnosis IgG4-RD and activity of the disease is the investigation of circulating plasma blasts but require flow cytometry facilities and are not disease-specific (Iaccarino et al., 2022).

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A simple CT scan with contrast is not sensitive enough to detect disease activity. We have good experience using PET/CT: we perform it routinely at the beginning of the diagnostic process, followed by a second examination after 6 months of therapy. Subsequently, we perform PET/CT as needed for each patient. In 5 patients, we did not detect metabolic activity in the infiltrate at the time of the examination, although 2 patients had signs of chronic renal insufficiency, and 1 patient had a unilaterally functional kidney. We did not start drug therapy in all of these patients, and the disease did not become active over the next few years. Therefore, it proves that a small number of patients may experience spontaneous recovery of disease activity (Williams et al., 2013).

Negative parameters of inflammation correlated with inactivity demonstrated by PET/CT. The clinical symptoms are non-specific in patients with Ormond's disease. There is no specific laboratory parameter for diagnosis or a biomarker to predict disease recurrence. The responsiveness of the disease to systemic corticosteroid therapy with a decrease in inflammatory parameters can be considered to confirm the diagnosis. Differentiating Ormond's disease from oncological disease is sometimes problematic (Sica et al., 2019). Hence, if possible, a biopsy should be performed to confirm the diagnosis. If the biopsy under CT guidance does not lead to an unequivocal verification and there are clinical doubts about the nature of the disease, then it is necessary to perform an operative revision and collect a representative sample for histological verification. We have repeatedly dealt with a situation where a patient was sent to us with suspicion of RF and a biopsy proved they had cancer.

From the point of view of laboratory parameters, Ormond's disease is characterised by positivity for inflammatory markers. However, these parameters are non-specific. Imaging methods (CT and PET/CT) provide great help in the diagnosis (Peisen et al., 2020). IRF can manifest as chronic periaortitis. This nosological unit describes several diseases with inflammatory involvement of the aorta – IRF, inflammatory aneurysm of the abdominal aorta, and perianeurysmal RF. We described inflammatory aneurysm of the abdominal aorta as part of IgG4-RD several years ago (Laco et al., 2013; Prucha et al., 2019). In these patients, however, the resulting abdominal aortic aneurysm is primarily treated surgically due to the practical impossibility of performing a biopsy and subsequent histological examination. It is worth noting that in the past 12 years, when we have been following patients with abdominal aortic aneurysm, there has not been a recurrence of the disease in any case after surgery. Perianeurysmal RF manifests as dilatation of the aorta and the presence of a fibrotic process that affects the adjacent organs (e.g., the ureters).

**Therapy**

There is not a consensus regarding how to treat Ormond's disease (Vaglio and Maritati, 2016; Tanaka and Masumori, 2020; Gianfreda et al., 2023; Vianello et al., 2023). The first-line treatment often involves systemic corticosteroids alone or in combination with other immunosuppressants. In our cohort, 42 patients received a combination of corticosteroids with azathioprine, and 41 patients received corticosteroids alone. The starting dose was not higher than 60 mg of prednisone or 48 mg of methylprednisolone. The patients received the starting dose for 12–16 weeks, followed by gradual tapering to 5 mg over 2 weeks. In the case of using azathioprine, we used a dose of 2×50 mg/day with a reduction after 3 months. In 7 patients, we stopped after 4–12 weeks due to elevated liver test results. The results of the therapy did not differ between the groups that did and did not receive azathioprine. When patients received corticosteroids and azathioprine, we were able to stop corticosteroids more quickly. On the other hand, long-term use of corticosteroids caused hyperglycemia in 27 patients, necessitating the initiation of oral antidiabetic drugs. In 16 patients, these drugs could be discontinued after the therapy ended.

Out of the 83 patients, 10 (12%) experienced disease exacerbation between 7 and 24 months after the discontinuation of immunosuppressive treatment. This result aligns with the findings of Kermani et al. (2011). Mycophenolate mofetil therapy was successfully used in 5 patients with disease exacerbations.

We treated 3 patients with repeated exacerbations and systemic complications (IgG4-RD) using rituximab. For all three patients, we implemented a treatment regimen that included maintenance therapy six months after initiation. None of the three patients experienced disease exacerbation during the two years following the start of treatment. Additionally, no significant hypogammaglobulinemia occurred in any of these patients. Rituximab is currently considered safer and more effective than corticosteroids, taking into account their side effects.

A Boston study included 26 patients with retroperitoneal fibrosis (Wallwork et al., 2018). Of these, 19 were evaluated as retroperitoneal fibrosis associated with IgG4-RD, while 7 were classified as idiopathic retroperitoneal fibrosis. All patients were treated with rituximab. A total of 19 (73%) received rituximab as monotherapy, while the rest received it in combination with glucocorticoids.

Radiographic evaluations were conducted in 25 patients, with 22 (88%) showing radiographic improvement (reduction in infiltrate size). Among the 10 patients with ureteral stents and/or percutaneous nephrostomy, stents or nephrostomies could be removed in 4 (40%). This study confirmed the effectiveness of rituximab monotherapy for patients with retroperitoneal fibrosis, regardless of whether the fibrosis was associated with IgG4-RD or idiopathic. However, the study does not provide long-term follow-up data post-treatment.

The second retrospective study evaluating the benefits of rituximab specifically for patients with idiopathic retroperitoneal fibrosis was published in Canada (Boyeva et al., 2020). In all ten patients, regression of fibrotic infiltrate size was confirmed via imaging studies. However, the authors of this study highlighted unresolved questions. While maintenance therapy with rituximab prevents relapses, the optimal duration of therapy remains unclear.

A nationwide French study analysed 156 patients registered in their database, of whom 33 were treated with rituximab (Ebbo et al., 2017). A clinical therapeutic response was observed in 29 out of 31 evaluated patients (93.5%). During a median follow-up of 24.8 months, the disease relapsed in 13 out of 31 patients with therapeutic response, with a median interval of 19 months from the last rituximab application. This study was the first to evaluate maintenance therapy and demonstrated the absence of relapses with consistent use of maintenance therapy. Therefore, the authors strongly recommend maintenance therapy, emphasizing its benefits despite the increased risk of infections due to rituximab-associated hypogammaglobulinemia. However, the optimal duration of this therapy remains an open question.

Following the success of rituximab in treating lymphoproliferative diseases, other anti-CD20 monoclonal antibodies, such as ofatumumab and obinutuzumab, have been introduced. As of 2024, only one publication describes the use of ofatumumab in retroperitoneal fibrosis (Hanazono et al., 2022).

## Conclusion

Ormond's disease (IRF) involves serious complications including renal insufficiency or failure and aneurysm of the abdominal aorta or iliac arteries. Its diagnosis and treatment are based on an interdisciplinary approach. In recent years, the possibilities of therapy have expanded significantly, which had improved the quality of life of patients and has reduced the development of serious complications.

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# Morphometry of Iliac Bones – A Useful Guide for Harvesting Bone Grafts

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**Key words:** Ilium – Hip bone morphometry – Iliac crest thickness – Orthopedic reconstructive surgery

**Abstract:** Iliac crest is common site for harvesting bone grafts. Morphometry of iliac crest is of vital importance in orthopedic surgery. Measurements were done on male (n=85) and female (n=85) hip bones. Length of iliac crest, thickness of iliac crest and ilium were measured. Thickness was measured at pre-defined points on crest and ilium 2 cm apart starting from anterior superior iliac spine (ASIS). Ilium was measured at a depth of 2.5 cm from crest. Statistical analysis was done. Iliac crests were longer in male bones. Ventral iliac crest was thickest at 6 cm from ASIS in both sexes. While iliac crest bore minimum thickness at 12 cm and 10 cm from ASIS in male and female bones respectively, however at 2.5 cm below iliac crest surface ilium was thickest at 4 cm from ASIS and at ASIS in male and female bones respectively. In case of male bones, dorsal part of iliac crest was thickest at 2.15 ± 1.29 cm from posterior superior iliac spine (PSIS) while in females it was at 1.78 ± 1.31 cm from PSIS. In dorsal part of ilium, it was observed at 2.31 ± 1.47 cm and 1.9 ± 1.79 cm from PSIS for male and female bones respectively. This study provided detailed variable morphometry and significant sexual dimorphism observed in iliac crest and ilium. Thickest safe zones in both sexes are a useful guide for harvesting appropriate bone grafts.

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## Introduction

The iliac bones of patients play an important role as one of the best sources of autologous bone graft. Morphometry of iliac bones would help orthopedic and spine surgeons to plan and harvest different types of bone graft as per indication, whether a good tricortical graft for interbody spinal fusion or adequate quantity of cancellous and cortico-cancellous grafts for filling in bone defects or for treating non-union of fractures of long bones (Grotz et al., 2005; Balogh et al., 2007; Garden et al., 2012).

Bone graft may be harvested during surgery with the patient in supine and sometimes lateral position or spine surgeons may need to harvest iliac crest graft in prone position, hence morphometry of the entire ilium from anterior to the posterior end is necessary.

Historically morphometry of the iliac crest has been explored but in isolated aspects (Xu et al., 1996; Ebraheim et al., 1997; Mahato, 2011). This study was designed to provide the surgeon with comprehensive quantitative data on length and thickness of the entire adult iliac crest and ilium with special emphasis on the variability in these parameters between sex and laterality. This study presents thorough information to orthopedic surgeons to determine thickness of different sites of the iliac crest for harvesting appropriate size of tricortical iliac bone grafts specially for anterior cervical discectomy and fusion (ACDF) with patient in supine for anterior approach of cervical spine or anterior interbody fusion of thoracic spine by transthoracic approach in lateral position or for harvesting slivers of cortico-cancellous bones for posterior spinal fusion in prone position.

## Material and Methods

Study was conducted on 170 dry human hip bones present in the osteological museum of the Department of Anatomy, Maulana Azad Medical College, New Delhi. While 83 bones were of right side comprising of 43 male and 40 female iliac bones, 87 bones were of left side comprising of 42 male and 45 female iliac bones. Thus, there were equal number of male and female iliac bones, 85 from each sex. Data concerning body height, age, pelvic width, and sex of iliac bones was not available. Sex of iliac bones was determined using metrical parameters like chilotic line, acetabulum-pubis index, and non-metrical parameters like ischiopubic rami and pre-auricular sulcus (Tubbs, 2016). Length of iliac crest was measured from anterior to posterior superior iliac spine (ASIS, PSIS).

To measure the thickness of iliac crest, ASIS was identified and thereafter points were marked at a regular interval of two cm starting from the ASIS to PSIS both on the surface of iliac crest as well as 2.5 cm below iliac crest (i.e. in the upper third of ilium) (Figure 1).

The thickness of iliac crest and ilium was measured at these defined points using micrometer/screw gauge and mean, standard deviation and range were calculated from data. All parameters were measured by two researchers in isolated settings. Each individual

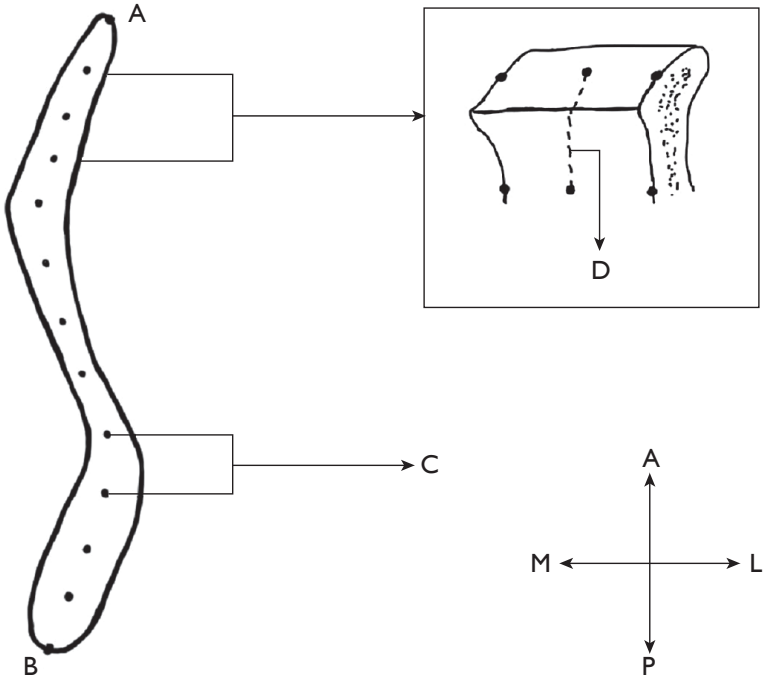


*Figure 1: Schematic diagram of superior view of left hip bone to show points marked on iliac crest at regular intervals of two cm starting from anterior superior iliac spine (ASIS) to posterior superior iliac spine (PSIS). Inset shows points marked on ilium 2.5 cm below iliac crest to measure its thickness. A – ASIS; B – PSIS; A–B – length of iliac crest; C – predefined points two cm apart on iliac crest where thickness of iliac crest was measured; D – predefined point 2.5 cm from point on iliac crest where thickness of ilium was measured.*

**Table 1: Length of iliac crest (mean, SD, and range)**

| | Total | Male | Male (R) | Male (L) | Female | Female (R) | Female (L) |
|---|---|---|---|---|---|---|---|
| Mean length ± SD (in cm) | 20.84 ± 2.05 | 21.92 ± 1.46 | 21.43 ± 1.49 | 22.42 ± 1.25 | 19.75 ± 1.99 | 19.98 ± 1.76 | 19.56 ± 2.17 |
| Range max–min (in cm) | 25.5–14.5 | 25.5–19 | 25.5–19 | 25–20 | 23–14.5 | 22.5–15.5 | 23–14.5 |

SD – standard deviation; R – right; L – left; p-value for male and female is < 0.0001, for male R and L is 0.01, for female R and L is 0.33

did measurements thrice and mean was recorded as final value.

The thickness of iliac crest and ilium at different sites were compared for any statistically significant difference between two sexes, as well as for variation among right and left bones in either sex, using independent sample *t*-test (SPSS version 17 [trial version]). P-value < 0.05 was taken as significant.

This study was conducted with prior ethical approval from the Institutional Ethics Committee. All local and international ethical guidelines and laws that pertain to the use of human bones in anatomical research were followed.

## Results

The length of iliac crest measured from ASIS to PSIS is depicted in Table 1 with maximum, minimum and mean length of iliac crests in both sexes. Length of iliac crest in males was greater than that of females and the difference was statistically significant (p-value < 0.001). Left male iliac crest (mean: 22.42 ± 1.25 cm) was significantly longer than right (21.43 ± 1.49 cm) (p-value = 0.01). Within female hip bone cohort however right iliac crest (19.98 ± 1.76 cm) was longer than left (19.56 ± 2.17 cm) but not significantly.

The mean thickness of iliac crest and ilium at the predefined points in both male and female hip bones is tabulated in Table 2 (Figure 2). In both male and female hip bones, site of maximum thickness of iliac crest was found to be located six cm from ASIS, on both sides. Average thickness of male iliac crests was significantly (p-value < 0.001) greater than female iliac crests at this location. The mean thickness of thinnest portion of iliac crests in males was 0.85 ± 0.15 cm. However, thinnest point on male iliac crests differed among right and left sides at different locations. Ventral segment of male iliac crests was thinnest on left side, located 12 cm from ASIS (mean value = 0.84 ± 0.15), while on right side it was thinnest at ten cm distance (mean value = 0.86 ± 0.22 cm). In female iliac crests, thinnest site was located ten cm from the ASIS (mean value of 0.74 ± 0.14 cm) symmetrically on both sides. At thinnest point too, male iliac crests were significantly thicker than female iliac crests (p-value < 0.001).

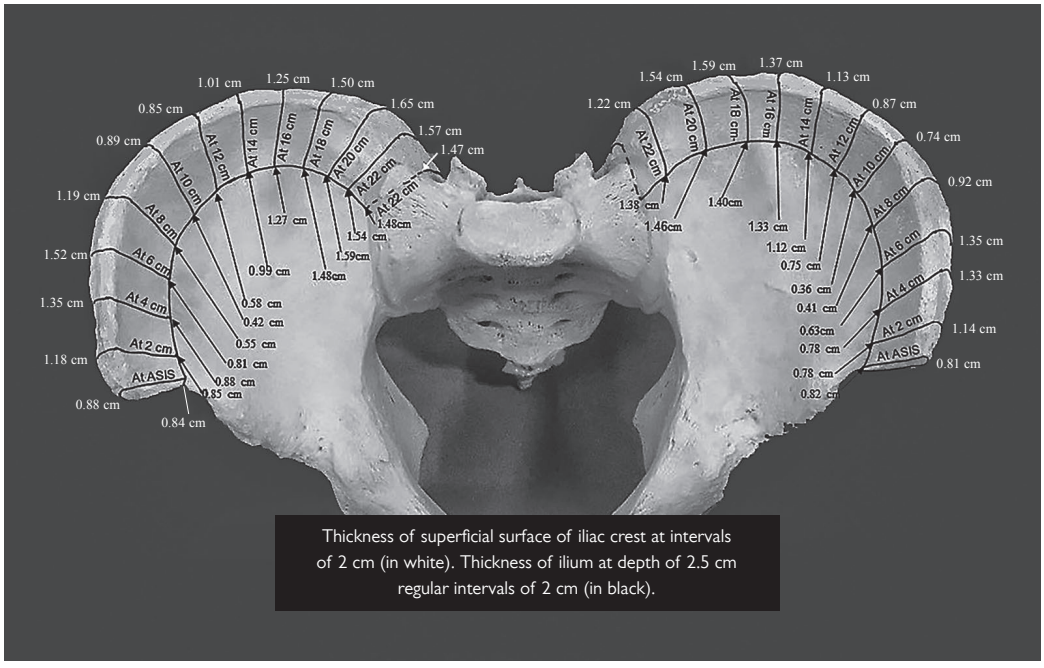


*Figure 2: Pictorial representation of thickness of iliac crest (in white) and ilium (in black) in males (left) and females (right) at intervals of 2 cm.*

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**Table 2: Mean thickness of iliac crest and ilium at the predefined specific points**

| Distance from ASIS (in cm) | Mean thickness of iliac crest with SD (in cm), n=85 | | P-value | Mean thickness of male ilium with SD (in cm), n=85 | | P-value |
|---|---|---|---|---|---|---|
| | male | female | | male | female | |
| 0 | 0.88 ± 0.18 | 0.81 ± 0.20 | **0.030** | 0.84 ± 0.17 | 0.82 ± 0.16 | 0.350 |
| 2 | 1.18 ± 0.19 | 1.14 ± 0.19 | 0.190 | 0.85 ± 0.14 | 0.78 ± 0.15 | **0.010** |
| 4 | 1.35 ± 0.26 | 1.33 ± 0.22 | 0.650 | 0.88 ± 0.20 | 0.78 ± 0.21 | **0.010** |
| 6 | 1.52 ± 0.24 | 1.35 ± 0.29 | **<0.001** | 0.81 ± 0.25 | 0.63 ± 0.25 | **<0.001** |
| 8 | 1.19 ± 0.33 | 0.92 ± 0.23 | **<0.001** | 0.55 ± 0.27 | 0.41 ± 0.21 | **0.010** |
| 10 | 0.89 ± 0.23 | 0.74 ± 0.14 | **<0.001** | 0.42 ± 0.23 | 0.36 ± 0.19 | 0.100 |
| 12 | 0.85 ± 0.15 | 0.87 ± 0.20 | 0.600 | 0.58 ± 0.35 | 0.75 ± 0.40 | **0.010** |
| 14 | 1.01 ± 0.25 | 1.13 ± 0.23 | **0.010** | 0.99 ± 0.39 | 1.12 ± 0.33 | **0.030** |
| 16 | 1.25 ± 0.27 | 1.37 ± 0.23 | **0.010** | 1.27 ± 0.31 | 1.33 ± 0.24 | 0.100 |
| 18 | 1.50 ± 0.33 | 1.59 ± 0.31 | 0.070 | 1.48 ± 0.31 | 1.40 ± 0.25 | 0.090 |
| 20 | 1.65 ± 0.35 | 1.54 ± 0.35 | 0.070 | 1.59 ± 0.31 | 1.46 ± 0.31 | **0.020** |
| 22 | 1.57 ± 0.36 | 1.22 ± 0.39 | **0.010** | 1.54 ± 0.32 | 1.38 ± 0.37 | 0.110 |
| 24 | 1.47 ± 0.51 | | | 1.48 ± 0.31 | | |

ASIS – anterior superior iliac spine; SD – standard deviation

Difference in thickness of ilium between male and female bones was statistically significant at 2, 4, 6, 8, 12, 14 and 20 cm (Table 2). In male hip bones, ventral part of ilium was thickest at 4 cm from ASIS with mean of 0.88 ± 0.20 cm. Values however showed a wide range between 0.36 and 1.47 cm. On the other hand, in female hip bones thickest point was just beneath ASIS (zero cm) with mean thickness of 0.82 ± 0.16 cm while thinnest part of the ilium was at 10 cm from ASIS. Values ranged between 0.07 and 1.08 cm with mean value of 0.39 ± 0.21 cm. Surprisingly, this point was similar for entire cohort. Mean value of thinnest dimension in male ilium was 0.42 ± 0.23 cm, while in female ilium it was 0.36 ± 0.19 cm. This difference was not statistically significant (p-value = 0.1). In 12 bones, five males and seven females, ilium thickness was less than or equal to 0.1 cm at ten cm from ASIS.

Dorsal segment of iliac crest was thickest at 2.15 ± 1.29 cm from the PSIS in case of male hip bones. Values ranged from maximum of 2.62 cm to minimum of 1.18 cm with value of 1.82 ± 0.28 cm on an average. However, in female iliac bones, the site of maximum thickness was located 1.78 ± 1.31 cm from PSIS, with mean value of 1.67 ± 0.29 cm. Difference between two sexes was statistically significant (p-value < 0.001) (Table 3).

In dorsal part of ilium, maximum thickness of male hip bones was observed at 2.31 ± 1.47 cm from PSIS with mean value of 1.74 ± 0.28 cm. Values ranged between 1.16 and 2.42 cm. Male hip bones were significantly thicker than female hip bones (p-value = 0.01). However, thickest site for female hip bones was located at 1.9 ± 1.79 cm from PSIS. Difference between two sides was not statistically significant in any subset (Table 4).

Table 4 provides comparative measurements of the thickness of iliac crest and ilium of the right and left sides. In both male and female hip bones, the thickness was variably more on right than left side, although these values were not statistically significant.

## Discussion

Before puberty and adolescent spurt of growth there is almost no difference between hip bones of boys and girls. Characteristic changes of shape, angulation and thickness occur after puberty, with male bones becoming thicker than female bones (Bryce, 1915; Crockford and Converse, 1972). Literature however, states that there are no statistically significant differences between various morphometric parameters in male and female hip bones (Mahato, 2011). In contrast, we observed some significant differences between the two sexes on extensive mapping of 170 dry adult hip bones.

Zaker Shahrak et al. (2014) measured the length of iliac crest as 24.75 cm with a range of 21.95 to 27.36 cm with male iliac crests being significantly longer than female. Similarly, we too found a variation in length of iliac crest between male and female,

**Table 3: Mean thickness of iliac crest and ilium at the predefined specific points**

| Distance from ASIS (in cm) | Mean thickness of male iliac crest with SD (in cm), n=85 | | P-value | Mean thickness of female iliac crest with SD (in cm), n=85 | | P-value |
|---|---|---|---|---|---|---|
| | right | left | | right | left | |
| 0 | 0.91 ± 0.18 | 0.84 ± 0.18 | 0.06 | 0.84 ± 0.18 | 0.79 ± 0.20 | 0.20 |
| 2 | 1.18 ± 0.20 | 1.17 ± 0.18 | 0.91 | 1.16 ± 0.20 | 1.12 ± 0.17 | 0.38 |
| 4 | 1.36 ± 0.25 | 1.34 ± 0.27 | 0.63 | 1.36 ± 0.23 | 1.31 ± 0.22 | 0.32 |
| 6 | 1.49 ± 0.24 | 1.56 ± 0.24 | 0.16 | 1.36 ± 0.30 | 1.35 ± 0.28 | 0.87 |
| 8 | 1.10 ± 0.30 | 1.28 ± 0.34 | 0.06 | 0.91 ± 0.21 | 0.93 ± 0.26 | 0.68 |
| 10 | 0.86 ± 0.22 | 0.93 ± 0.23 | 0.16 | 0.74 ± 0.14 | 0.75 ± 0.14 | 0.82 |
| 12 | 0.87 ± 0.15 | 0.84 ± 0.15 | 0.35 | 0.88 ± 0.22 | 0.86 ± 0.17 | 0.60 |
| 14 | 1.08 ± 0.24 | 0.94 ± 0.24 | 0.06 | 1.15 ± 0.27 | 1.12 ± 0.20 | 0.48 |
| 16 | 1.27 ± 0.27 | 1.23 ± 0.27 | 0.46 | 1.37 ± 0.24 | 1.38 ± 0.23 | 0.94 |
| 18 | 1.56 ± 0.37 | 1.44 ± 0.28 | 0.10 | 1.58 ± 0.32 | 1.60 ± 0.29 | 0.70 |
| 20 | 1.65 ± 0.40 | 1.66 ± 0.30 | 0.95 | 1.60 ± 0.35 | 1.49 ± 0.36 | 0.25 |
| 22 | 1.55 ± 0.38 | 1.60 ± 0.34 | 0.58 | 1.11 ± 0.38 | 1.38 ± 0.35 | 0.13 |
| 24 | 1.29 ± 0.48 | 1.56 ± 0.54 | 0.48 | | | |

| Distance from ASIS (in cm) | Mean thickness of male ilium with SD (in cm), n=85 | | P-value | Mean thickness of female ilium with SD (in cm), n=85 | | P-value |
|---|---|---|---|---|---|---|
| | right | left | | right | left | |
| 0 | 0.84 ± 0.19 | 0.84 ± 0.14 | 0.89 | 0.79 ± 0.15 | 0.85 ± 0.17 | 0.09 |
| 2 | 0.86 ± 0.14 | 0.83 ± 0.14 | 0.33 | 0.79 ± 0.12 | 0.78 ± 0.18 | 0.81 |
| 4 | 0.88 ± 0.17 | 0.88 ± 0.23 | 0.95 | 0.79 ± 0.21 | 0.77 ± 0.21 | 0.75 |
| 6 | 0.75 ± 0.23 | 0.86 ± 0.25 | 0.04 | 0.63 ± 0.27 | 0.63 ± 0.23 | 0.99 |
| 8 | 0.50 ± 0.23 | 0.60 ± 0.31 | 0.12 | 0.42 ± 0.24 | 0.39 ± 0.19 | 0.60 |
| 10 | 0.43 ± 0.23 | 0.40 ± 0.23 | 0.11 | 0.37 ± 0.20 | 0.36 ± 0.18 | 0.87 |
| 12 | 0.68 ± 0.40 | 0.48 ± 0.24 | 0.12 | 0.70 ± 0.39 | 0.79 ± 0.41 | 0.30 |
| 14 | 1.11 ± 0.43 | 0.86 ± 0.31 | 0.10 | 1.14 ± 0.33 | 1.10 ± 0.33 | 0.56 |
| 16 | 1.32 ± 0.31 | 1.21 ± 0.31 | 0.13 | 1.34 ± 0.24 | 1.33 ± 0.23 | 0.91 |
| 18 | 1.52 ± 0.34 | 1.43 ± 0.27 | 0.18 | 1.41 ± 0.27 | 1.40 ± 0.24 | 0.90 |
| 20 | 1.56 ± 0.32 | 1.62 ± 0.30 | 0.39 | 1.45 ± 0.33 | 1.46 ± 0.29 | 0.91 |
| 22 | 1.52 ± 0.33 | 1.56 ± 0.31 | 0.67 | 1.36 ± 0.42 | 1.43 ± 0.31 | 0.68 |
| 24 | 1.34 ± 0.09 | 1.53 ± 0.36 | 0.24 | | | |

ASIS – anterior superior iliac spine; SD – standard deviation

**Table 4: The average maximal thickness of the dorsal iliac crest and ilium in male and female hip bones of both sides along with standard deviation and range (as measured from the PSIS)**

| | Male | | | Female | | |
|---|---|---|---|---|---|---|
| | mean | | p-value | mean | | p-value |
| | right | left | | right | left | |
| **Dorsal iliac crest** | 1.84 ± 0.3 (range: 2.62–1.28) | 1.80 ± 0.28 (range: 2.26–1.81) | 0.54 | 1.72 ± 0.28 (range: 2.35–1.19) | 1.64 ± 0.30 (range: 2.11–0.88) | 0.21 |
| | mean | | p-value | mean | | p-value |
| | right | left | | right | left | |
| **Dorsal ilium** | 1.74 ± 0.28 (range: 2.42–1.19) | 1.73 ± 0.28 (range: 2.38–1.16) | 0.98 | 1.62 ± 0.2 (range: 2.05–1.26) | 1.52 ± 0.26 (range: 2.22–0.87) | 0.06 |

PSIS – posterior superior iliac spine

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however mean the iliac crest length in Indian subset is 20.84 ± 2.05 cm, much smaller than Caucasian crest. Depending on the type and/or purpose for which bone graft is needed, bone is obtained from either posterior ilium or anterior ilium or iliac crest. Ebraheim et al. (2001) reported that the ideal site for cortico-cancellous bone graft is either 3 cm posterior to ASIS or 6 cm anterior to PSIS, while tricortical iliac crest grafts are better obtained in supine position 3 cm posterior to ASIS. In a biomechanical analysis of iliac crest loading after bone graft harvest, Schmitz et al. (2018) emphasized the importance of iliac crest and iliac blade as sites of attachment of abdominal muscles, muscles of the back, glutes, and muscles of the thigh namely sartorius, rectus femoris and tensor fascia lata. Thus, removal of block of harvested bone impairs transmission of force produced by the contraction of these muscles along iliac crest and ilium which predisposes the bone to avulsion fractures. Hence the location from where the bone is harvested and the size is crucial to maintaining mechanical integrity of the bone.

Ebraheim et al. (1997) reported the thickness of ilium at 2 and 3 cm from ASIS and at iliac tubercle as 1.06 ± 0.18 cm, 1.17 ± 0.18 cm, and 1.69 ± 0.23 cm, respectively and the anterior point of iliac tubercle was measured to be situated 3 cm posterior to ASIS. In the present study, the maximum thickness of anterior iliac crest was observed at 6 cm from ASIS in both sexes. The value was 1.52 ± 0.24 cm and 1.35 ± 0.29 cm in male and female bones respectively. Compared to our study, the dimensions reported by Ebraheim et al. (1997) are more possibly due to Caucasian study set and they advocated that bone graft is best taken from iliac tubercle where the bone was thickest and found no statistical difference with reference to sex or side. Contrary to this we have found statistically significant difference between the two sexes. We too propose that best site for taking bone graft is from around the iliac tubercle. Hwang et al. (1997) mapped 58 iliac bones for surgeons to localize the site of harvesting and also observed that the maximum thickness of ventral segment of iliac crest and ilium was present between ASIS and iliac tubercle. Although male hip bones were thicker in comparison to female in their study, difference was not statistically significant except at a few sites in the intermediate part near the crest (Hwang et al., 1997). Hu et al. (1995) based on their biomechanical analysis advocated that harvesting bone graft from ilium at ASIS or just 1.5 cm posterior to it resulted in decreased stability of the crest. The same when done 3 cm behind, preserves the strength 2.4 times more. Ropars et al. (2014) also postulated that safe distance from ASIS for harvesting was 2 cm posterior to it and an ideal graft length should not be more than 47 mm and must be from anterior to a line passing through the thickest part of iliac tubercle. The authors also compared their dissected specimen results with computed tomographic analysis and found no difference between the two. More recently Schmitz et al. (2018) also corroborated the fact that harvesting bone at a distance of 2–2.5 cm posterior to the ASIS would also suffice to reduce risk of fractures. They also concluded that it is advisable to maintain integrity of the crest and extract bone from ilium.

Mahato (2011), advocates that the ideal site of bone harvesting in Indian population is anterior to iliac tubercle in the ventral segment of iliac crest. He observed that the area of ilium located more than 45 mm from ASIS is the widest and contains the maximum thickness and volume of cancellous bone. He reported that the amount of bone in female ilium was lesser but not significantly so and proposed that harvesting should be done 3 cm posterior to the ASIS both in males and females.

On the basis of morphometric thickness of iliac crest and ilium measured in the present study we hypothesize that the optimum area to obtain cortico-cancellous grafts in Indian population varies depending on the sex. In male population ilium bone is thickest (0.88 ± 0.2 cm) at 4 cm posterior to ASIS; at 2 cm posterior to ASIS bone thickness is 0.85 ± 0.14 cm and at 6 cm bone is 0.81 ± 0.25 cm and thereafter thins out to 0.55 ± 0.27 cm at 8 cm and is the thinnest at 10 cm with mean value of 0.42 ± 0.23 cm. In fact, in 12 bones in our study sample, 5 males and 7 females, the ilium was papery thin (less than or equal to 0.1 cm) at 10 cm from ASIS. It is advisable therefore to harvest bone between 2 and 4 cm posterior to ASIS in males which can be easily extended to 6 cm if a greater length of the bone is needed. The thickness of the iliac crest varies from 1.18 ± 0.19 at two cm, 1.35 ± 0.26 at four cm to the thickest dimension of 1.52 ± 0.24 cm at six cm. Thus, “thickest safe zone” of iliac crest for harvesting bone grafts in male hip bones is 2–6 cm posterior to the ASIS.

On the other hand, in female hip bones, ilium is thickest immediately beneath ASIS. Harvesting bone at this site would however predisposes to stress fractures (Hwang et al., 1997; Mason et al., 2005). The next thickest segment of ilium is located at 2 and 4 cm (0.78 ± 0.15 and 0.78 ± 0.21) from ASIS. Iliac blades subsequently thin out substantially at six cm (0.63 ± 0.25 cm), eight cm (0.41 ± 0.21 cm) and is thinnest at ten cm (0.36 ± 0.19 cm). “Thickest safe zone” in females is more anteriorly placed as compared to males and is located between two cm to four cm posterior to ASIS. Thickness of iliac crest at two cm and four cm posterior to ASIS in females is 1.14 ± 0.19 cm and 1.33 ± 0.22 cm, although it is

thickest at six cm posterior to ASIS (1.35 ± 0.29 cm). Thus, our data suggests that safe zone for harvesting the ilium is anterior to the iliac tubercle in males and females and is two–four cm posterior to ASIS, extendable to six cm in males but not in females. Surgeons need to exercise caution since safe zones appear similar, yet there is a significant difference in thickness of ilium between males and females.

Posterior ilium is preferred site for bone graft harvesting especially when patient position during surgery is prone and is associated with less mortalities (Ropars et al., 2014). Xu et al. (1996) measured thickness of posterior iliac blade in six cadavers and 30 dry bones and categorized it into three zones. They reported zone 1 to be the safest to obtain graft with average thickness reported as 1.71 ± 0.22 cm. This zone extended along the posterior iliac crest from PSIS to the apex of sacroiliac articulation. The authors have documented a maximum of 2.2 cm and a minimum thickness of 1.4 cm in this zone. The thickness surprisingly for the dorsal segment was lesser than our Indian population. In the present study, the thickest region of posterior iliac crest in male bones is located at 2.15 ± 1.29 cm from PSIS (mean value: 1.82 ± 0.28 cm). Thus, the zone of maximum thickness in our study in male iliac crests was from 0.86 cm anterior to PSIS to 3.44 cm anterior to PSIS. Similarly, the thickest zone for dorsal segment ilium was on an average at 2.31 ± 1.47 cm anterior to PSIS with a mean thickness of 1.74 ± 0.28 cm. Thus, the thickest zone was from 0.84 to 3.78 cm anterior to PSIS. In female ilium this point was present at 1.9 ± 1.79 cm from PSIS having a mean thickness of 1.56 ± 0.24 cm. Female iliac crest was thickest at 1.78 ± 1.31 cm from PSIS (mean value: 1.67 ± 0.29 cm). The zone of maximum thickness in female iliac crest was from 1.38 to 1.96 cm anterior to PSIS. This zone for female ilium was from 1.32 to 1.8 cm anterior to PSIS. Again, dorsal segment of iliac crest and ilium were also significantly thicker in males as compared to females (p-value: < 0.001 and 0.01, respectively).

The thickest part of the bone needs to be identified to avoid cortical table penetration (Schmitz et al., 2018). As per our findings, the thickest portion of iliac blade was from ASIS till four–five cm posterior to it at a depth of 2.5 cm from the crest.

Lack of comparison between different age groups was one of the limitations of our study. Another difficulty which we encountered while comparing data was that thickness of ilium measured by different authors was at different depths whereas as the strength of this study is meticulous morphometric mapping at frequent intervals of the entire iliac bone.

## Conclusion

In Indian population, a significant morphometric dimorphism is noted among male and female pelvic bones. Iliac crest was significantly longer and thicker in male, though point of maximal iliac crest thickness (six cm posterior to ASIS) was same in both sexes. In both male and females, the ventral ilium is thickest, anterior to the thickest part of iliac crest. Based on the observations of the present study, for taking bone graft from ventral ilium, the point of harvesting should be anterior to the thickest part of iliac crest, at a safe zone of two–four cm posterior to ASIS, extendable to six cm in males, but not in females. If a larger size graft is needed in females, then other bones like fibula should be considered. The thickest part of anterior ilium for females is just beneath the ASIS, which is not an ideal site for harvesting bone graft. Dorsal segment of iliac crest and ilium are also significantly thicker in males as compared to females.

With the exhaustive data on iliac bone offered by the present study, surgeons should be able to select appropriate site of ilium and harvest appropriate dimension of bone graft as per their indication.

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# The Use of Front Plateau in the Treatment of Temporomandibular Disorders: A Case Series and Literature Review

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**Key words:** Temporomandibular joint – Temporomandibular joint disorders – Temporomandibular joint disc – Occlusal splints – Conservative treatment

**Abstract:** Temporomandibular disorders (TMDs) are conditions with multifactorial etiology and complex treatment. Among the non-invasive therapeutic possibilities for these conditions is the Front Plateau, a partial anterior plate made from colourless self-curing acrylic resin. It is a simple procedure that can be carried out in a single clinical section promoting muscle relaxation to reduce symptoms associated with TMDs. This study aims to report a prospective, consecutive, single-centric case series to evaluate the Front Plateau's effectiveness in patients with temporomandibular disorders. A questionnaire adapted from the Research Diagnostic Criteria for TMDs was used and 4 patients were treated with the Front Plateau plaque. Patients were monitored after 5 and 9 months, respectively, after starting to use the Front Plateau. Of the 4 cases listed, 2 showed significant improvement in initial signs and symptoms. Front Plateau may be a favourable treatment option for patients with TMD, if the guidelines are followed. Clinical trials on this modality should seek to minimize possible biases and limitations associated with the design of this type of research.

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## Introduction

Temporomandibular disorders (TMDs) are a set of signs and symptoms that may be of joint and/or muscle origin and are associated with masticatory muscles, the temporomandibular joint (TMJ), bone tissues, and adjacent soft tissues (Melchior et al., 2019; Eriksen et al., 2020). These conditions are among the most common pains of non-dental origin, with an overall prevalence of approximately 31%, and represent a relevant public health problem (Valesan et al., 2021). According to Blanco-Hungria et al. (2016), women between 30 and 40 years old are the group most affected by these conditions.

Known for their complex and multifactorial etiology, TMDs can be categorized into two axes: axis I, associated with physical conditions and related to nociceptive stimuli, and axis II, characterized by psychological factors that can cause or influence the pain experience of patients (Okeson, 2013; Schiffman et al., 2014; González-Sánchez et al., 2023). Among the etiological factors that can be listed as TMDs, are anatomical, occlusal, neuromuscular, psychological, hormonal, trauma, and/or parafunctional habits such as bruxism, tooth clenching, and nail-biting, which may be influenced by the emotional state of the patient (Cruz et al., 2020).

Generally, the signs and symptoms reported by individuals who suffer from TMDs include pain on palpation, joint noises and cracks, tension-type headaches, otalgia, pain in the face region, muscle fatigue, spasms, a change in mandibular trajectory during mouth opening and closing, bruxism, and tooth sensitivity due to wear (Felício and Braga, 2005; Sassi et al., 2018). Therefore, therapeutic approaches involve making rigid occlusal splints (total or partial), guidelines for patients with TMDs, laser therapy, physiotherapy, pharmacotherapy, and acupuncture, among other approaches that may or may not be associated (Piozzi and Lopes, 2002; Al-Moraissi et al., 2020).

In this perspective, a type of treatment that could be adopted in patients with complaints related to TMD is the confection of a partial anterior plate, commonly made with colourless self-curing acrylic resin and known as Front Plateau. Its confection must be carried out considering the anterior guides in the protrusion movements and the canine guide in the lateral movements, with disocclusion of the posterior teeth (Dekon et al., 2007). It is a technique indicated for a short period of time due to the possibility of extrusion of the posterior teeth since they do not touch during the use of the plaque (Okeson, 2013).

Although Front Plateau does not have a well-understood mechanism of action, the literature associates its effectiveness with muscle relaxation obtained through modification of proprioception, occlusion, vertical dimension, musculoskeletal stabilization, and the placebo effect (Oliveira, 2002). Although some reports show advantages over the use of the Front Plateau, there is still a lack of studies about its use as a therapeutic modality for patients with TMDs (Gomes et al., 2018; Belchior et al., 2021). Therefore, the objective of this study was to present a prospective, consecutive, and unicentric series of cases where the Front Plateau was used as a treatment for patients with TMD.

## Front Plateau: What is it, what is it for and why use it?

It is very common to find studies comparing occlusal splints with different therapeutic modalities. However, few studies evaluate the effectiveness of different occlusal devices against each other (Pficer et al., 2017; Al-Moraissi et al., 2022). The evaluation of the performance of therapeutic modalities by clinical trials can suffer from the Hawthorne effect, where participants tend to behave differently when they are taking part in a research study (Sedgwick and Greenwood, 2015). This can lead some researchers to associate the positive effect of occlusal splints with a placebo effect. However, a systematic review with meta-analysis revealed that the action of occlusal splints goes beyond this influence (Alkhutari et al., 2021).

Partial-coverage occlusal splints have emerged as an alternative to rigid full-coverage occlusal splints, which can involve a series of clinical steps, requiring more clinical time (Januzzi et al., 2010; Al-Moraissi et al., 2020; Alkhutari et al., 2021). In this context, the Front Plateau and a prefabricated device used by some professionals, the nociceptive trigeminal inhibition tension suppression system (NTI-tss), are possibilities reported in the literature (Jokstad, 2009). The main mechanism of action of both is the disocclusion of the posterior teeth, thus relieving the hyperactivity of the chewing muscles and protecting the teeth (Klasser and Greene, 2009). However, unlike the Front Plateau, which involves a larger number of teeth, the NTI-tss is small and has a greater risk of being aspirated by the patient, as reported in some studies. In addition, there is a need for a greater number of adjustments in the mouth (Jokstad et al., 2005; Stapelmann and Turp, 2008).

Therefore, the Front Plateau can be indicated and has been used by some researchers, thanks to its easy manufacture in a single session and low cost (Doepel et al., 2012; Gomes et al., 2018). In addition, it is possible to find in the literature the use

of the device to establish differential diagnoses of conditions associated with TMDs that are difficult to identify (Belchior et al., 2021). In a study of 20 patients comparing the use of the Front Plateau with a rigid full-coverage occlusal splint to control TMDs, it was found that there was no significant difference between the devices. Evaluations were conducted using electromyography and showed that both devices appear to be effective in controlling muscle pain associated with TMDs (Dahlström and Haraldson, 1985). These data corroborate another crossover clinical trial conducted with 71 patients, comparing patient satisfaction with the use of a full-coverage splint without the occlusal portion (placebo), the Front Plateau and a rigid full-coverage splint (Greene and Laskin, 1972).

Because this device only covers the anterior teeth, its use is limited to short periods of time. This is because, when used for prolonged periods of time, they can induce extrusion of the posterior teeth due to the lack of contact and mobility of the anterior teeth due to the accumulation of load during the use of the Front Plateau (Jokstad et al., 2005; Klasser and Greene, 2009).

## Material and Methods

This case series was prepared at the TMD clinic and conducted in accordance with the Declaration of Helsinki of 1975, revised in 2013 (World Medical Association, 2013). All participants provided informed consent forms. The anamnesis, clinical examination, diagnostic impressions, and proposed therapeutic modalities were carried out according to the clinical form adopted at the institution. Mouth opening characteristics and the presence of pain on palpation in muscle and joint regions were recorded according to a questionnaire adapted from the Research Diagnostic Criteria for Temporomandibular Disorders (RDC/TMD).

Therefore, the pain reported by the patients was classified on a scale from 0 to 3, where 0 – no pain, 1 – mild pain, 2 – moderate pain, and 3 – severe pain. After 5 months of starting to use the interocclusal devices, the patients were contacted via telephone to follow-up on the evolution of the treatment. This case series was developed following the preferred reporting of case series in surgery – PROCESS guideline (Agha et al., 2020).

To manufacturing the Front Plateau, some materials are needed: liquid and powder of self-curing acrylic resin; measuring pot; Vaseline; Paladon glass pot; carbon paper; milling cutters for finishing and discs for polishing; straight nosecone handpiece and low-speed handpiece; Spatula n° 31; Muller forceps; clinical kit. After assembling the work table, the powder and liquid of the self-curing acrylic resin are handling. Then, the resin in its rubbery phase is adapted to the patient's teeth (Figure 1A and B). After the material has completely cure, it is necessary to make occlusal adjustments (Figure 2A and B). After final adjustments, finishing and polishing (Figure 3A and B) the Front Plateau is ready.

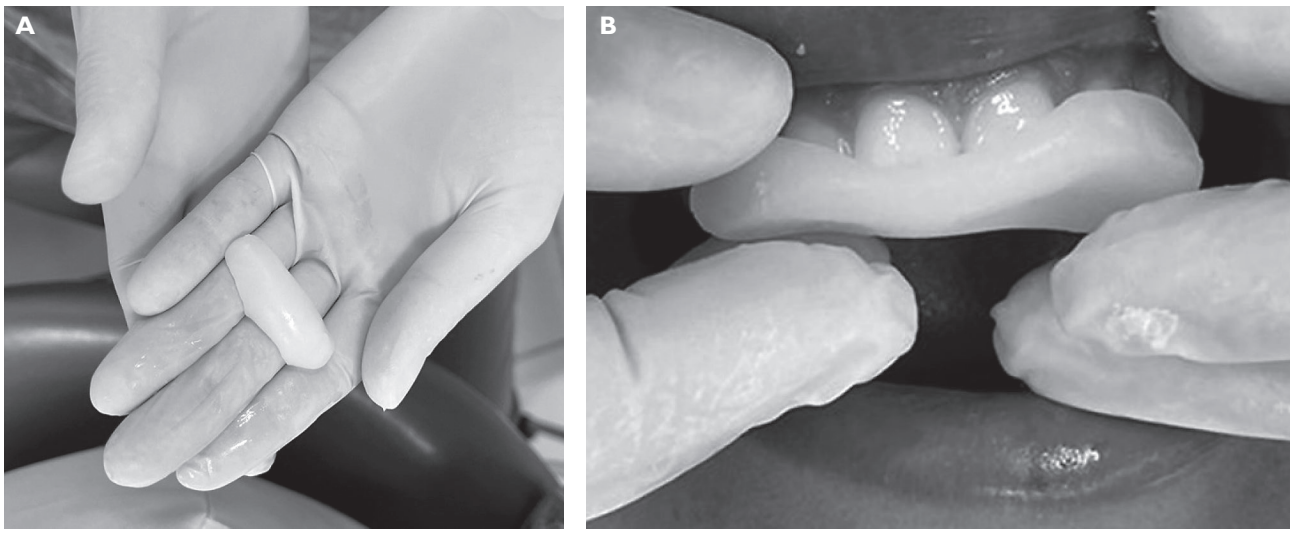


*Figure 1: Adaptation of the polyacrylic resin to the patient's teeth. A) After reaching its rubber phase, the patient's teeth were isolated with liquid Vaseline and the resin was removed from the Paladon glass pot and manipulated to obtain a format similar to that shown in the image. B) Then, the resin was adapted to the patient's upper anterior teeth. To allow free manipulation of the material, the operator rubbed Vaseline on the fingers so that the material would not stick. In addition, at some moments, the patients were instructed to close their teeth so that they could make small marks where the lower anterior teeth would fit. To avoid excessive heating during the setting of the material, throughout the entire process, the resin was removed from the patient's mouth and immersed in water.*

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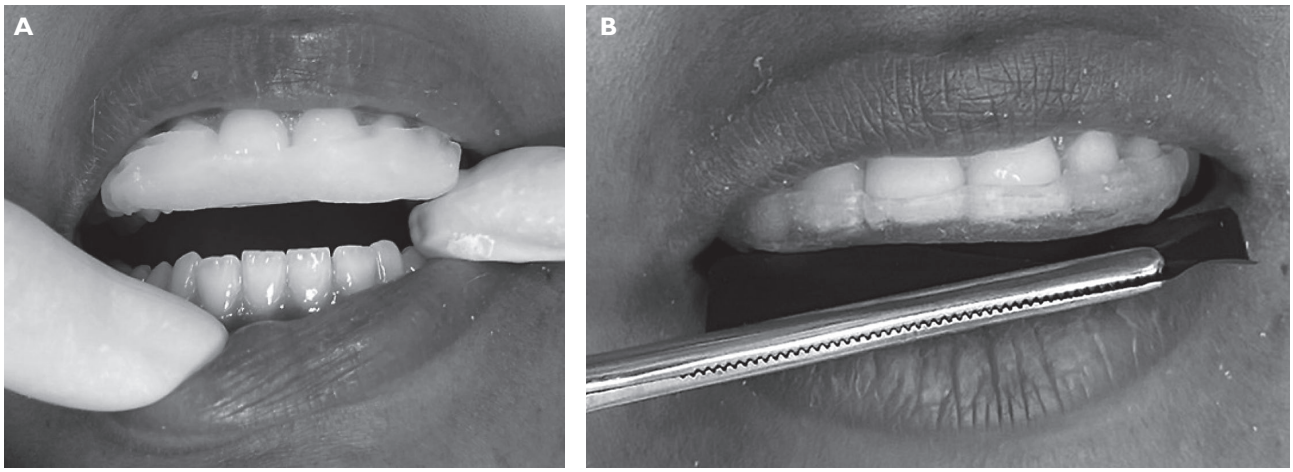


*Figure 2: Checking the contacts between the teeth. A, B) After setting the material, the contact points between the upper and lower anterior teeth were checked, assessing the need for adjustments before finishing and final polishing.*

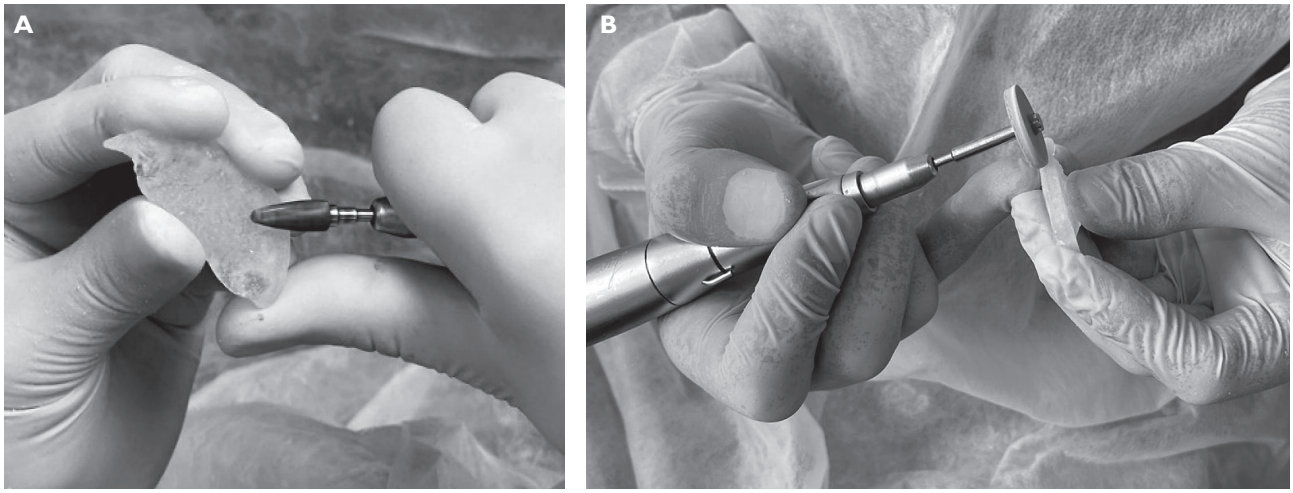


*Figure 3: Necessary adjustments, finishing and final polishing. A) When necessary, adjustments were made to the Front Plateau so that all the upper and lower anterior teeth touched each other during its use. B) Then, the final finishing and polishing were carried out with abrasive discs using a straight nosecone handpiece.*

## Cases description

### Case 1

A 26-year-old female was referred by the Otorhinolaryngologist with the following report: frequent otalgia on the left ear, without signs of otitis, with spontaneous improvement. Audiometry showed no alterations. Mild pain on palpation of the left TMJ and slightly limited mouth opening. The patient was using Otosylase 10,000 IU, a drug with anti-inflammatory and analgesic actions indicated to treat external ear inflammation. During the clinical examination, a difference between the centric relation (CR) and the maximum habitual intercuspation (MHI) greater than 4 mm and bilateral clicking during mouth opening were detected, being symmetrical and with soft active movement. The patient had the habit of grinding and clenching her teeth, chewing gum frequently, and continuously using the telephone. In addition, her head was slightly deviated to the left and had an Angle Class III facial pattern (postural).

When performing the palpation exam in different areas of the face, head, and neck, the patient reported grade 3 pain in the region of the masseter, lateral pterygoid, and temporalis tendon on the right and left sides. Among the diagnostic impressions were disc displacement with reduction and myalgia (local muscle soreness). In the same session, a low-power laser was applied following the manufacturer's recommendations (Laser Smile, Quick Smile) in these regions, and recommendations were also given regarding nighttime habits for sleep hygiene. In the following session, the Front Plateau plaque was made to stabilize the mandible in a more comfortable position for the TMJ

in centric relation. The patient was instructed to use the device at night.

Two weeks later, during re-evaluation, the patient reported a significant improvement in pain symptoms. Therefore, she was instructed to continue using the device overnight and was also referred to the Orthodontist to assess the need for orthognathic surgery or conventional orthodontic treatment. In the reassessment appointment five months later, the patient reported no symptoms except pain when the weather was colder, requiring the use of Otosylase 10,000 IU. A new evaluation was carried out 9 months after using the Front Plateau and the patient reported the absence of initial symptoms, except on colder days.

**Case 2**

A 57-year-old female sought care complaining of pain and clicking in the TMJ region. According to the patient, the pain started after traumatic personal events four years prior. She reported having depression, fibromyalgia, bruxism, and teeth clenching and using medications such as Pregabalin (75 mg), Duloxetine (30 mg), Trazodone (50 mg), Losartan, Pantoprazole (40 mg) and Tramadol (50 mg).

When performing the palpation examination in different areas of the face, head, and neck, the patient reported grade 2 pain in the region of the temporalis, middle pterygoid, and suprahyoid muscles on the right and left sides. In the region of the lateral pterygoid muscles and temporalis tendon and the TMJ area (lateral pole and posterior ligament) on both sides, grade 3 pain was reported. Among the diagnostic impressions were disc displacement with reduction, myalgia (local muscle soreness) and joint hypermobility. In the same session, low-power laser therapy was applied according to the manufacturer's recommendations (Laser Smile, Quick Smile) in these regions, and the Front Plateau plaque was made in CR to stabilize the mandible in a more comfortable position for the TMJ, relaxing the facial muscles. The patient was informed about the recommendations and guidelines for patients with TMD and stress management.

After one week of, the patient returned to the clinic reporting that the Front Plateau plaque was "not well adapted and loose" and that the muscle and joint pain had not improved. The plaque was adjusted, and a new low-power laser therapy application was performed in the same regions previously described. In the following week, the patient reported a slight improvement in the symptoms of muscle and joint pain, but the discomfort persisted.

The patient was referred to receive a total rigid myorelaxant plaque. When reassessed after 5 months of using the Front Plateau, there was a partial reduction in the clicks, but the pain persisted. According to the patient, on days with acute fibromyalgia symptoms, the pain was strong, but on other days it was milder. A new assessment was carried out 9 months after using the Front Plateau and the patient reported the same situation observed in the 5-month assessment.

**Case 3**

A 49-year-old female sought care complaining of pain and clicking in the TMJ. According to the patient, she had suffered a fall in 2010, and in 2015, the discomfort started as a headache. She used Propranolol (40 mg), Levothyroxine (25 mg), Sertraline (50 mg), and Amitriptyline (25 mg) and had the habits of biting her nails, continuous use of the telephone, and reported being talkative. During clinical examination, she reported painful symptoms when performing mouth movements of maximum opening, protrusion, and right and left laterality, in addition to opening with deviation and joint noise on the left side.

When performing the palpation examination in different areas of the face, head, and neck, the patient reported grade 3 pain in the region of the temporalis, masseter, lateral pterygoid muscles, temporalis tendon, and in the TMJ region (lateral pole and posterior ligament) of both sides. Among the diagnostic impressions, disc displacement with reduction, myalgia (local muscle soreness), and a tension-type headache stood out.

In the same session, a low-power laser therapy was applied according to the manufacturer's recommendations (Laser Smile, Quick Smile) in these regions. In addition, guidelines for patients with TMDs were given, and a referral was made for evaluation and treatment by a physiotherapist.

After a week, the patient returned to the clinic reporting persistent pain. In view of the case, it was proposed to make a Front Plateau plaque as an attempt to treat the discomfort, aiming to observe whether there would be an indication for a total rigid myorelaxant plaque afterwards. Two months later, the patient returned to the clinic to assess the evolution of the treatment. A significant improvement in pain on the right side was reported, with grade 3 pain persisting only in the lateral pterygoid area. Regarding the left side, there was a decrease in joint noise and improvement in pain only in the temporalis muscle, with persistent grade 3 pain in the other regions evaluated by palpation. The patient reported performing physiotherapy on the left side due to the persistence of pain in this region of the face; however, due to the significant improvement presented on the right side during the use of the Front Plateau plaque,

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she was referred to receive a total rigid myorelaxant plaque.

In the reassessment 5 months later, the patient reported a considerable reduction in the pain symptoms on both sides, but in some periods, she observed that the pain returned, however, at a lower intensity than it was before the use of the Front Plateau plaque. A new evaluation was carried out 9 months after starting to use the Front Plateau and the patient reported an improvement in initial symptoms but an increase in pain after carrying out activities that require physical effort.

**Case 4**

A 28-year-old female sought care complaining of severe pain in the TMJ region. According to the patient, the pain had started four years prior. She used Topiranate (25 mg) and Venlafaxine hydrochloride (150 mg) and had a habit of clenching her teeth and continuous use of the telephone and computer, in addition to being talkative. During the clinical examination, the patient reported that she was under stress during the period in which the first consultation was held.

When performing the palpation exam in different areas of the face, head, and neck, the patient reported grade 2 pain in the region of the temporalis muscles on the left side, in the masseter on both sides, and in the TMJ region (lateral pole) on both sides. On palpation of the region of the lateral pterygoid muscles and temporalis tendon, the patient reported grade 3 pain on the right and left sides. Among the diagnostic impressions, myalgia (local muscle pain) and myositis stood out. In the same session, a low-power laser therapy was applied according to the manufacturer's recommendations (Laser Smile, Quick Smile) in these regions. Additionally, the patient received orientations for patients with TMDs.

One week after the first consultation, the patient returned to the clinic and reported an improvement in the pain. Another session of low-power laser therapy was performed in the lateral pterygoid muscle and temporalis tendon regions. In the following week, despite the improvement in the patient's pain, the patient reported less intense discomfort; therefore, a Front Plateau was made. Upon returning for reassessment two weeks later, the patient reported complete remission of the painful symptoms, demonstrating interest in maintaining the use of the Front Plateau plaque since, according to her, it brought a great improvement in her quality of life.

In the reassessment after 5 months of using the Front Plateau plaque, there were no symptoms. Due to the patient's anxiety, her doctor prescribed

**Table 1: Clinical characteristics and evolution of patients after treatment**

| Patient | Main complaint | Risk factors | Medication already in use | Diagnostic impressions | Conduct prior to treatment | Post-treatment assessment |
|---|---|---|---|---|---|---|
| Case 1 | Otalgia and pain in the left TMJ<br>Limitation of mouth opening | Difference between MHI and CR > 4 mm | Otosylase 10,000 UI | Localized myalgia and DDWR | Guidelines for TMD patients | 5/9 months:<br>No symptoms with the use of the FP<br>Pain only on colder days |
| Case 2 | Pain and clicking in the TMJ region | Depression, fibromyalgi, bruxism, and tooth clenching | Donaren 50 mg<br>Duloxetine 30 mg<br>Losartana 50 mg<br>Pantoprazole 40 mg<br>Pregabalina 75 mg<br>Tramadol 50 mg | Localized myalgia, DDWR and joint hypermobility | Guidelines for TMD patients | 5 months: Partial reduction of symptoms<br>9 months: Further reduction in symptoms<br>Pain still very strong in acute periods of fibromyalgia |
| Case 3 | Pain and clicking in the TMJ region | Onychophagia, continuous cell phone use, and talking a lot | Amitriptilin 25 mg<br>Levotiroxina 25 mg<br>Propanolol 40 mg<br>Sertralina 50 mg | Localized myalgia, DDWR, and tension-type headache | Guidelines for TMD patients | 5/9 months:<br>Considerable reduction in symptoms<br>Pain worsens after activities that require physical exertion |
| Case 4 | Pain in the TMJ region | Teeth clenching, talking a lot, continuous use of cell phone and computer | Pregabalin 75 mg<br>Topiramate 50 mg<br>Venlafaxine 75 mg | Localized myalgia and myositis | Guidelines for TMD patients | 5/9 months:<br>No symptoms with the use of the FP |

Source: own authorship. TMJ – temporomandibular joint; DDWR – disc displacement with reduction; TMD – temporomandibular dysfunction; FP – Front Plateau; MHI – maximum habitual intercuspation; CR – centric relationship

Venlaxine (150 mg) and Pregabalin (150 mg). A new evaluation was carried out 9 months after starting to use the Front Plateau and the patient reported the absence of initial symptoms. A general overview of the cases can be seen in Table 1.

## Discussion

All reported cases are female; this fact can be justified by epidemiological surveys that show a higher prevalence of TMD in adult women, requiring therapeutic interventions (Medeiros et al., 2011; Blanco-Hungria et al., 2016). Among the four patients who received the Front Plateau plaque, two reported significant improvement in pain symptoms with the use of the anterior partial plaque following the orientations (Cases 1 and 4). The evolution of the cases was monitored for a period of 5 months and a new evaluation of the patients was carried out after 9 months of using the Front Plateau, it was noticed that the balance and reduction of muscular hyperactivity promoted by the Front Plateau plaque contributed to the reduction of discomfort reported during clinical examination (Molina, 1989; Dekon et al., 2007).

Another indication for the use of anterior partial plates is their emergency use when it is not possible to make an immediate total rigid plaque (Alencar et al., 1998; Oliveira, 2002). This was observed in the two patients who reported little or no improvement in the pain associated with TMDs (Cases 2 and 3), who were then referred to continue treatment. Another relevant aspect of the cases concerns the variety of diagnostic impressions listed for each patient, a fact that highlights the multifactorial characteristics of TMDs (Cruz et al., 2020).

In this perspective, initially less invasive interventions were adopted, such as low-power laser therapy for biostimulation and analgesic effects, to reduce the inflammatory and painful processes (Nadershah et al., 2020). In addition, orientations and exercises were given according to the condition of each patient to reduce the complaints presented, eliminate parafunctional and predisposing habits to TMDs, and stimulate a more harmonious relationship of the stomatognathic system (Oliveira et al., 2015; Resende, 2019). These interventions were adopted because the predisposing factors for TMD reported by patients, when not modified, hinder the success of treatment associated with interocclusal devices (Madani and Mirmortazavi, 2011; Chan et al., 2022). In cases of patients with severe pain, the modification of behavioural and psychological factors must be adopted from the first consultation and maintained even after the use of interocclusal splints (Garstka et al., 2023).

There are studies in the literature (Jokstad et al., 2005; Al-Moraissi et al., 2022) that point to the possibility of occlusal changes due to the use of anterior partial devices such as the Front Plateau and the NTI-tss. However, a cross-sectional study (Araújo et al., 2021) did not find a statistically significant relationship between the use of an anterior partial interocclusal device and tooth movements. However, due to its larger size, the Front Plateau plaque offers greater security than the NTI-tss, which has a smaller size and poses a risk of ingestion (Al-Moraissi et al., 2022). Another advantage presented by the Front Plateau is its low cost and the fact that its manufacture can be performed by the dentist in the dental office directly on the patient's dentition (Gomes et al., 2018; Belchior et al., 2021).

Despite not providing solid evidence, the present study contributes to future clinical trials on the use of the Front Plateau plaque for the treatment of TMD. Because it is a series of cases that describe outcomes with different patients, it is an appropriate study to verify adverse effects and intercurrences associated with the device (Grimes and Schulz, 2002). Case series, despite presenting a low level of scientific evidence, are important studies for the construction of scientific knowledge. This study design aims to bring benefits to clinical practice and can also generate notes and the possibility of further research, sometimes being the first source of evidence on a given subject (Oliveira et al., 2015).

One of the limitations of this study is that it does not compare groups since it does not have the methodological rigor observed in randomized clinical trials with a control group (Jüni et al., 2001; Polit and Gillespie, 2010). Furthermore, the patients were prospectively evaluated without comparing each other and according to the outcomes reported by them before and after the treatment.

The association with low-power laser therapy, even though it is a non-invasive treatment, may have influenced the results obtained (Assis et al., 2012). In addition, the patient in Case 3 had appointments with a physiotherapist, which may also have contributed to the relief of pain in the muscles involved (Silva Santos and Pereira, 2016). In addition to these factors, all patients were taking medications to control pain and muscle inflammation, as well as medications to control stress and anxiety (Cases 2, 3 and 4). This association with the use of Front Plateau is a major limiting factor, especially in the case of those used for stress, anxiety and depression, as they may be related to the emergence and/or worsening of TMD symptoms (Reissmann et al., 2012; Augusto et al., 2016; Florjański and Orzeszek, 2021).

In theory, a case series should provide descriptive information with sufficient detail to allow replication of the study by other researchers, thus enabling the creation of hypotheses and the discovery of innovative therapies. An example of this was a case series on COVID-19 published in January 2020 (Huang et al., 2020) which has supported several clinical trials and currently has a total of 30,381 citations by ScienceDirect and 54,973 by Google Scholar.

## Conclusion

Based on this case series, it was possible to observe that the Front Plateau plaque presents a favourable therapeutic approach for patients with TMDs when used for short periods of time and following the recommendations of the professional in charge. However, it is worth remembering that, as they are multifactorial, TMDs will often require multiprofessional treatment so that acute or chronic symptoms can be controlled. In the case of studies that aim to test the effectiveness of this treatment modality, possible biases associated with the research design, sample, and applied therapy must be considered and, when possible, minimized.

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# An Infective Endocarditis Case Report Involving Both Native Aortic and Mitral Valves Due to *Streptococcus Vestibularis*

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**Key words:** Infective endocarditis – *Streptococcus vestibularis* – Periannular abscesses – Bicuspid aortic valves

**Abstract:** Infective endocarditis (IE) is a life-threatening disease, with its mortality rate varying depending on the infectious agent. Streptococci are among the most common causes of infective endocarditis. However, *Streptococcus vestibularis* has rarely been associated with human infections, typically affecting patients with underlying conditions such as immunosuppressive diseases, valve replacement, rheumatic heart disease, and hemodialysis. We present the case of a 26-year-old man who presented with fever, unanticipated weight loss, and fatigue. Although no typical risk factors for infective endocarditis were identified at admission, transesophageal echocardiography revealed a bicuspid aortic valve with calcification, paravalvular aortic abscess formation, and vegetations on the anterior leaflet of the mitral valve. Blood cultures grew *S. vestibularis,* which was initially sensitive to benzylpenicillin but developed emergent resistance on the third day of the antibiotic treatment. Subsequently, ceftriaxone therapy was initiated, and blood cultures became sterile on day 10. The patient eventually underwent aortic valve replacement. We report the first known case of native aortic and mitral valve endocarditis caused by *S. vestibularis*, accompanied by a paravalvular abscess around the native aortic valve, in a patient who had no typical risk factors for infective endocarditis, except for a bicuspid aortic valve.

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## Introduction

Infective endocarditis (IE) is an infection of the endocardial layer of the heart, typically caused by bacteria or fungi. *Staphylococcus aureus* is responsible for approximately 31% of cases, primarily affecting the heart valves (both native and prosthetic). *Viridans streptococci* are the second most common cause, accounting for 17% of cases, while enterococci rank third in frequency (Hubers et al., 2020). Risk factors for infective endocarditis include intravenous drug use, the presence of heart valve diseases such as a bicuspid aortic valve, implanted intracardiac devices, and prosthetic heart valves (Vincent and Otto, 2018). Infective endocarditis can lead to systemic complications due to embolic events. It may progress to involve cardiac vegetations, valvular and periannular abscesses, and mycotic aneurysms (Hubers et al., 2020).

*Streptococcus vestibularis* was first isolated from the vestibular mucosa of human oral cavities and described as a new species of *Streptococcus viridans* in 1988 (Whiley and Hardie, 1988).

In a study on antibiotic resistance among Viridans group streptococci, 50 blood culture isolates were found to exhibit resistance to penicillin and tetracycline in 30% of cases, to clindamycin in 40%, and to cefotaxime in 20%. All isolates were susceptible to vancomycin, chloramphenicol, levofloxacin, and linezolid (Ergin et al., 2011). We aimed to present the first reported case of infective endocarditis involving both the native mitral and aortic valves due to *S. vestibularis* in the literature.

## Case report

A 26-year-old man was admitted to the hospital with a 2-month history of fever, fatigue, night sweats, and an unexpected weight loss of 10 kg. He had no known medical conditions or allergies, and this was his first hospital admission. His family history was unremarkable, aside from a paternal history of diabetes mellitus. He had been smoking cigarettes for 10 years.

Regarding risk factors for infective endocarditis, he had no history of intravenous drug use, rheumatic fever, or recent surgical or dental procedures. However, poor dental hygiene and dental caries were noted on physical examination. Cardiac auscultation revealed a mid-systolic murmur at the second intercostal space on the right sternal border, radiating to the neck, as well as a systolic murmur heard best at the apex, radiating to the left axilla. Otherwise, the physical examination was unremarkable, with no signs of infective endocarditis.

Abdominal examination revealed hepatosplenomegaly. Laboratory tests showed normocytic, normochromic anemia (hemoglobin level of 10 g/dl) and neutrophilic leukocytosis (white blood cell count of $15.2\times10^3$/µl, with 86% neutrophils). Serum analysis revealed normal vitamin B12 and folic acid levels, elevated ferritin, and decreased iron, consistent with anemia of chronic disease. Routine liver, thyroid, and kidney function tests were within normal limits. However, C-reactive protein (CRP) and erythrocyte sedimentation rate were elevated. The ASO (antistreptolysin O) test was negative.

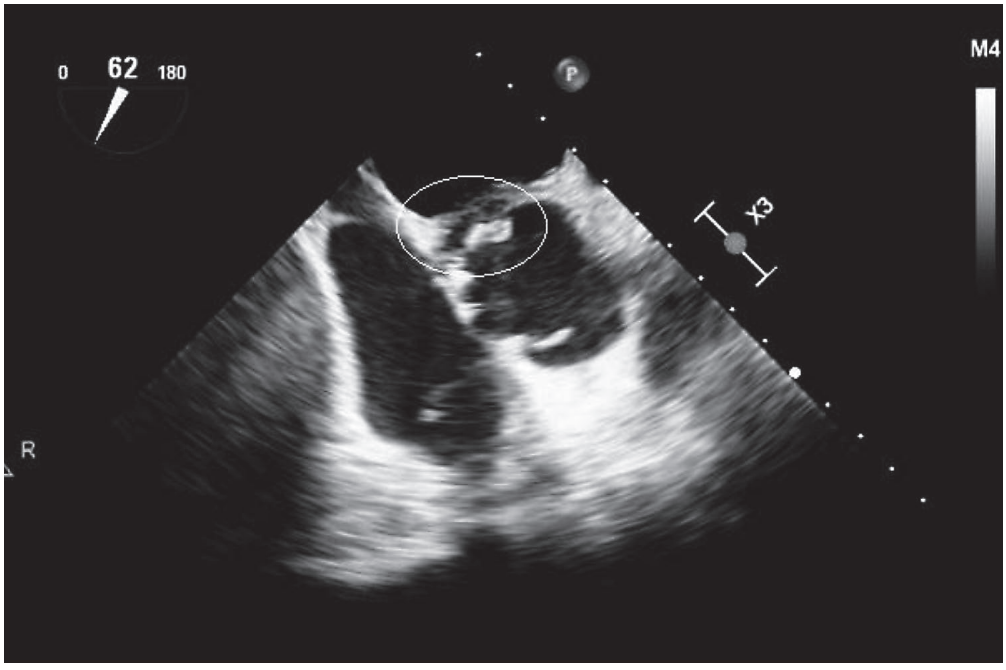


*Figure 1: Transesophageal echocardiography revealed the presence of a paravalvular abscess around the aortic valve, along with vegetations.*

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The first set of blood cultures (two bottles) grew *Streptococcus vestibularis*, which was sensitive to benzylpenicillin, and treatment with benzylpenicillin was initiated. Two additional sets of blood cultures were sent to the laboratory on the third and sixth days, both of which unexpectedly grew *S. vestibularis* resistant to benzylpenicillin. Consequently, antibiotic therapy was switched to ceftriaxone.

A thoracoabdominal computed tomography (CT) scan and a dental consultation were requested to investigate the origin of the *S. vestibularis*. The dental examination revealed no abnormalities apart from generally poor oral hygiene. The thoracoabdominal CT scan showed hepatosplenomegaly and hypodense lesions in the spleen, consistent with splenic infarction.

Transthoracic echocardiography revealed a bicuspid aortic valve with calcifications, accompanied by severe regurgitation and mild stenosis. Transesophageal echocardiography demonstrated a paravalvular abscess around the aortic valve and vegetations on the anterior leaflet of the mitral valve (Figure 1). No other complications of infective endocarditis were observed.

After 10 days of ceftriaxone therapy (2 g daily), blood cultures became sterile. The patient's clinical condition remained stable, with the exception of fluctuating fever and elevated C-reactive protein. Physical examination findings were unchanged. A second transesophageal echocardiogram on the 10$^{th}$ day revealed severe aortic regurgitation, along with persistent paravalvular abscess formation.

The patient subsequently underwent aortic valve replacement surgery without any perioperative complications. However, he experienced an ischemic stroke on the 7$^{th}$ postoperative day. Following medical and surgical treatment, the patient made a full recovery and was discharged on oral anticoagulation therapy.

## Discussion

Infective endocarditis has an increasing incidence and a high mortality rate worldwide. Reports indicate that 10,000 to 15,000 cases occur annually in the United States (Slipczuk et al., 2013). IE is associated with serious complications, prolonged hospitalization, surgery, long-term antibiotic therapy, significant economic burden, and high mortality (Vahabi et al., 2019; Chen et al., 2022). The average age of individuals diagnosed with IE has increased in the United States; however, the average age in Turkey remains lower at 47 years. The male-to-female ratio is approximately 1.5:1 (Slipczuk et al., 2013; Vahabi et al., 2019).

IE caused by *Streptococcus viridans* is typically associated with dental procedures. However, our patient had no history of any dental interventions and no other known risk factors for IE, aside from the bicuspid aortic valve, which was detected later. The most common etiological agents of IE include *Staphylococcus aureus*, *Streptococcus viridans*, enterococci, and coagulase-negative staphylococci, respectively (Hubers et al., 2020).

A review of the literature (PubMed, Google Scholar, Web of Science, Scopus, EMBASE, and EBSCO) using the keywords "infective endocarditis" and "*Streptococcus vestibularis*" identified five articles (five case reports), none of which described an instance of infective endocarditis involving both native aortic and mitral valves (Cunliffe and Jacob, 1997; Partridge, 2000; Doyuk et al., 2002; Tufan et al., 2010; Kuwauchi et al., 2023). To our knowledge, our case represents the first reported instance of IE caused by *S. vestibularis* in which both native aortic and mitral valves were affected.

*Streptococcus viridans* is a significant component of the human oropharyngeal microbiota and can cause serious infections, including infective endocarditis, abscesses, and bacteremia, particularly in neutropenic patients (Fish et al., 1995). The antibacterial activity of penicillin against *Streptococcus viridans* depends on its binding affinity to penicillin-binding proteins (PBPs). Penicillin resistance can occur when this affinity is reduced, typically due to alterations in one or more amino acids near the penicillin-binding site of the PBPs. While *S. viridans* isolated from IE cases are generally sensitive to penicillin, bacteremia caused by penicillin-resistant *S. viridans* is common in neutropenic patients (Lopardo et al., 2022). In our case, *S. vestibularis,* which was initially sensitive to benzylpenicillin, grew in the first two sets of blood cultures. However, it unexpectedly developed emergent resistance to benzylpenicillin in subsequent cultures. As a result, antibiotic therapy was switched to ceftriaxone.

Penicillins and aminoglycosides are the antibiotics most commonly associated with the development of resistance during monotherapy. In contrast, *Streptococcus* species have a 13% emergent resistance rate during antibiotic therapy with penicillin (Fish et al., 1995).

This patient had only one identified risk factor for IE: a bicuspid aortic valve. The source of *S. vestibularis* remains unclear, but the most likely origin is oral flora, given the patient's poor oral hygiene. Furthermore, no abnormal findings were detected during physical examination or thoracoabdominal computed tomography except hepatosplenomegaly and splenic infarcts, which could suggest a plausible source for

*S. vestibularis*. These splenic infarctions were consistent with and supported the diagnosis of IE.

This was the patient's first hospital admission. His clinical presentation was typical for IE; however, the involvement of both the aortic and mitral valves, the unusual causative agent (*S. vestibularis*), and the unclear source of infection were atypical features for IE.

## Conclusion

We report a case of infective endocarditis caused by *S. vestibularis*. To the best of our knowledge, this is the first reported case of IE due to *S. vestibularis* involving both native aortic and mitral valves, with an annular paravalvular abscess.

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# A Case of Pleuroparenchymal Fibroelastosis

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**Key words:** Pleuroparenchymal fibroelastosis – Postoperative – Weight loss – Prognosis

**Abstract:** Pleuroparenchymal fibroelastosis (PPFE) is a rare fibrotic lung disease with a poor prognosis. Some patients with PPFE have prominent lesions in one upper lobe. Due to the unilateral nature of lesions, similarities between some patients of PPFE and unilateral upper field pulmonary fibrosis (unilateral upper-PF) have been indicated. A 55-year-old man was referred to our hospital with PPFE, which had developed dyspnea 9 months previously. He had undergone resection of renal cancer 64 months prior to the onset of PPFE. He was administered the antifibrotic drug nintedanib, but passed away due to respiratory failure 43 months after the onset. What were impressive points in the clinical course of this PPFE patient was the similarity to unilateral upper-PF, including his medical history, and the poor prognosis. Although very rare, we do believe that the information on medical history and progression in this patient might provide suggestion into the treatment of future patients with a similar trajectory.

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## Introduction

Pleuroparenchymal fibroelastosis (PPFE) is a progressive disease that causes apical pleural thickening and pulmonary fibrosis in upper lobes of lungs. PPFE is a type of pulmonary fibrosis with a poor prognosis (Nakamura et al., 2021). Patients with PPFE include middle-aged and elderly men with a history of smoking (Nakamura et al., 2021). Although the etiology and pathology of this disease, like other pulmonary fibroses, remains unknown (Nakamura et al., 2021; Cottin et al., 2022), but it is known to occur in patients with a medical history of bone marrow or lung transplantation (Higo et al., 2019; Rasciti et al., 2019). Whereas some patients with PPFE have prominent lesions in one upper lobe. Due to the unilateral nature of lesions, similarities between some patients of PPFE and unilateral upper field pulmonary fibrosis (unilateral upper-PF) have been indicated (Sekine et al., 2017; Inafuku et al., 2022). Many patients with unilateral upper-PF have a history of thoracic surgery, and some studies have suggested

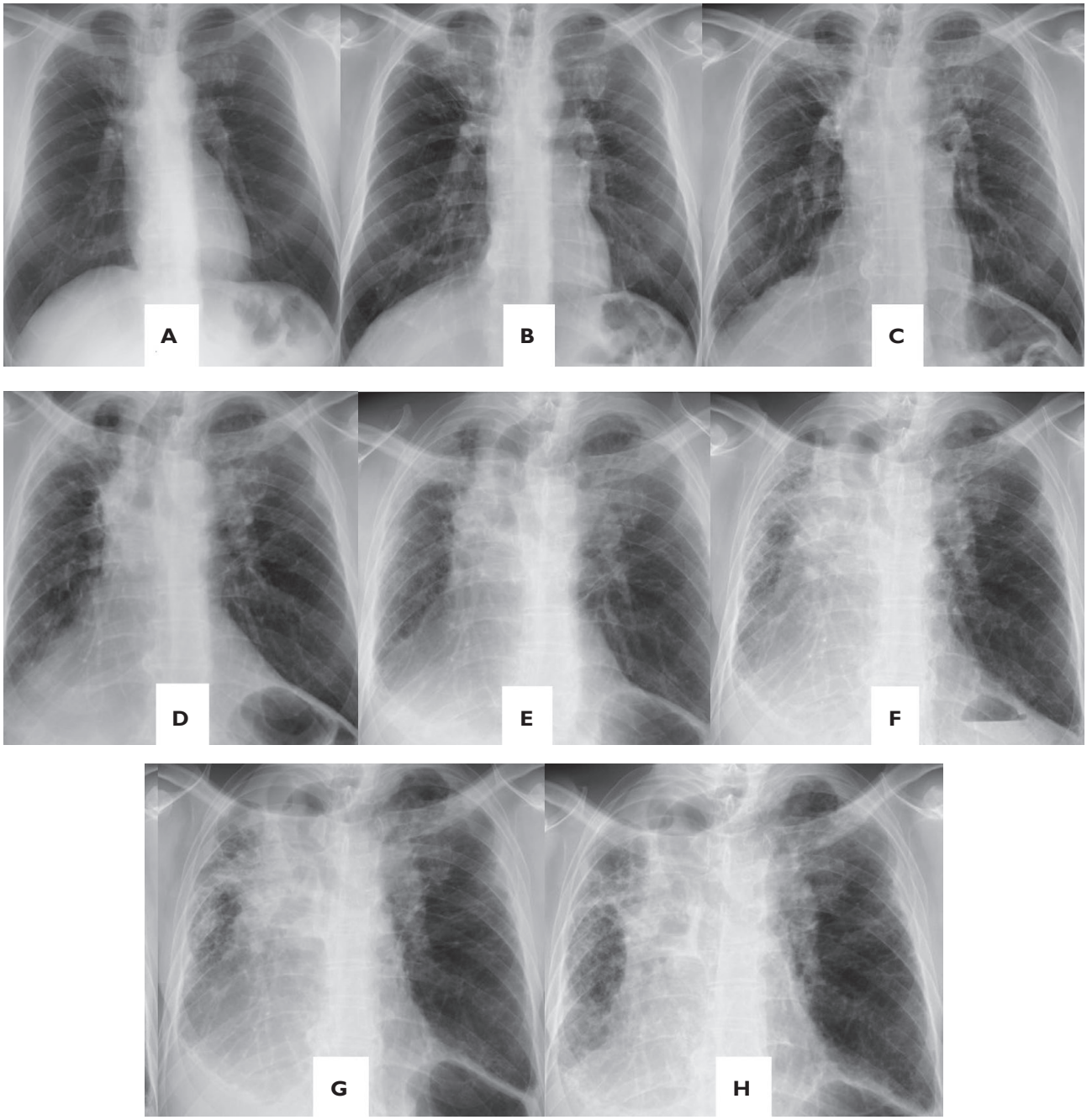


*Figure 1: Changes in chest radiographs over time. Chest radiograph taken just before resection of renal cancer 64 months before onset (A); chest radiograph taken 34 months before onset (B); chest radiograph taken 8 months before onset (C); chest radiograph taken at onset of pleuroparenchymal fibroelastosis (D); chest radiograph taken 8 months after onset (E); chest radiograph taken 18 months after onset (F); chest radiograph taken 25 months after onset (G); and chest radiograph taken 36 months after onset (H).*

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a relationship between the onset of fibrosis and this medical history (Sekine et al., 2017; Inafuku et al., 2022).

This time, we treated a patient with PPFE who had prominent lesions on the right side. This patient had no history of thoracic surgery, but had undergone surgical treatment for renal cancer 64 months before the onset of PPEF. Although the mechanism of onset of PPFE was unknown, we do believe that this clinical course would provide some suggestive information on treatment for patients with a similar course in the future.

## Case report

A 55-year-old man was referred to our hospital after moving. He had a 10-pack-year smoking habit but no history of occupational inhalation of particulate matter. Laparoscopic left nephrectomy for renal cell carcinoma (clear cell subtype, G1>G2, pT3a, V+) had been performed 64 months before the onset of respiratory symptoms, but a chest radiograph taken at the time of the surgery showed no abnormal findings. Preoperative forced vital capacity (FVC) was 4.17 l (108.3%) and forced expiratory volume in one second

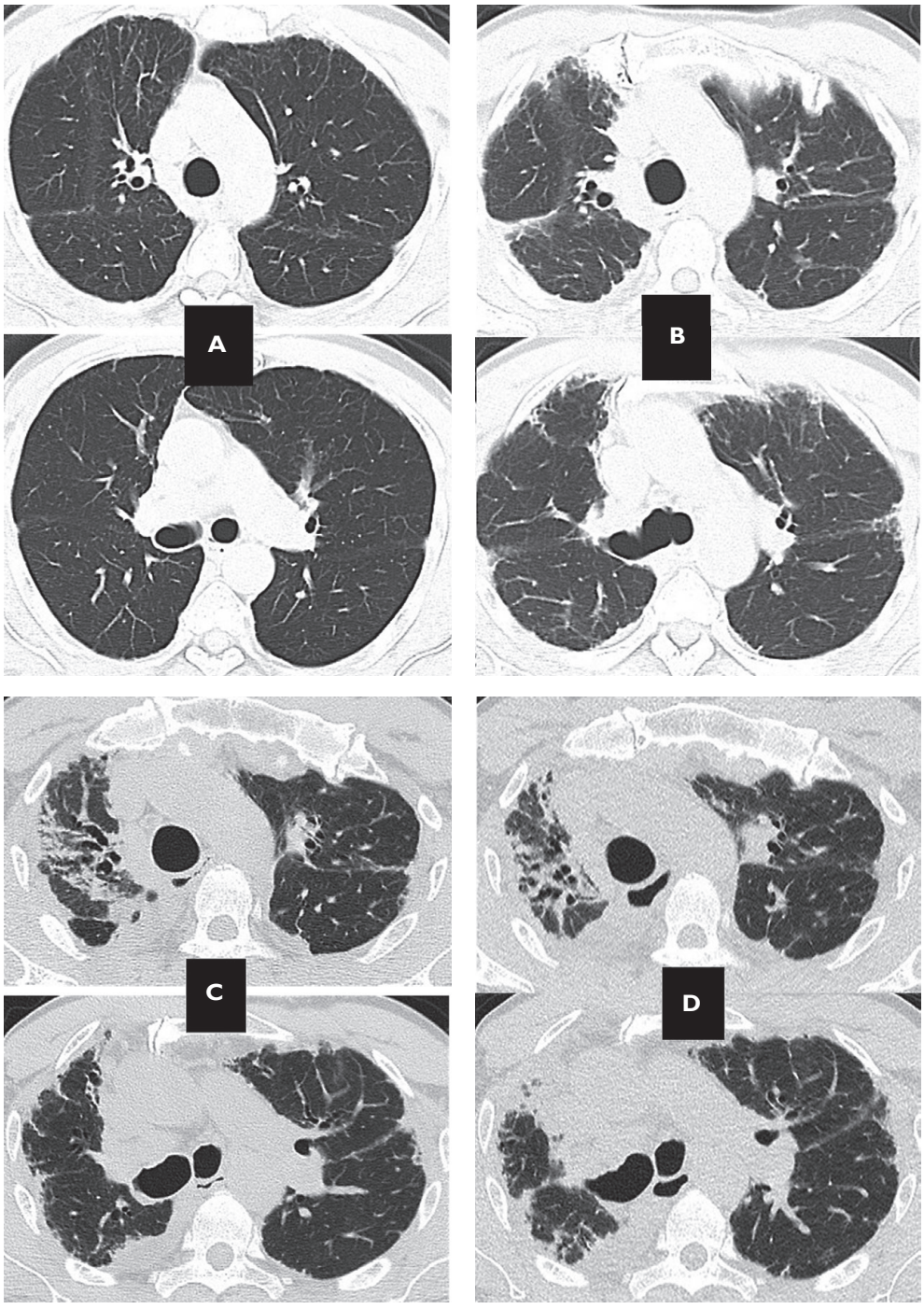


*Figure 2A: Changes in chest CTs (computed tomography) at the level of upper lobes and carina over time. Chest CT taken just before resection of renal cancer 64 months before onset (A); chest CT taken 6 months before onset (B); chest CT taken 9 months after onset (C); and chest CT taken 16 months after onset (D).*

($FEV_1$) 3.14 l (100.2%). There was no recurrence of the renal cancer thereafter, but approximately one year after the operation, the patient began to have dyspnea on exertion and dry cough. A chest radiograph taken around this time showed fibrosis predominantly in the upper lobes, with the lesion on the right side being prominent, and the shadow gradually intensified. Based on the radiological findings and clinical course, the patient was diagnosed with PPFE at another medical institution. When the patient was referred to our hospital 3 years after the onset of respiratory symptoms, he was thin, with a height of 163 cm, weight of 53 kg, and body mass index (BMI) of 19.9. Chest auscultation revealed no heart murmurs, but fine crackles were heard in both lung fields. At the time of his initial visit to our hospital, blood examination showed hemoglobin 14.6 g/dl, platelet count of 263,000/μl, albumin l 4.0 g/l, creatine 1.09 mg/dl, lactate dehydrogenase 184 U/l (124–222 U/l) and C-reactive protein (CRP) level of 0.10 mg/dl. Respiratory function test showed FVC 2.16 l (58.7%) and $FEV_1$ 1.98 l (64.3%), confirming a significant decrease in FVC. Two years after onset, the antifibrotic drug nintedanib was initiated, but

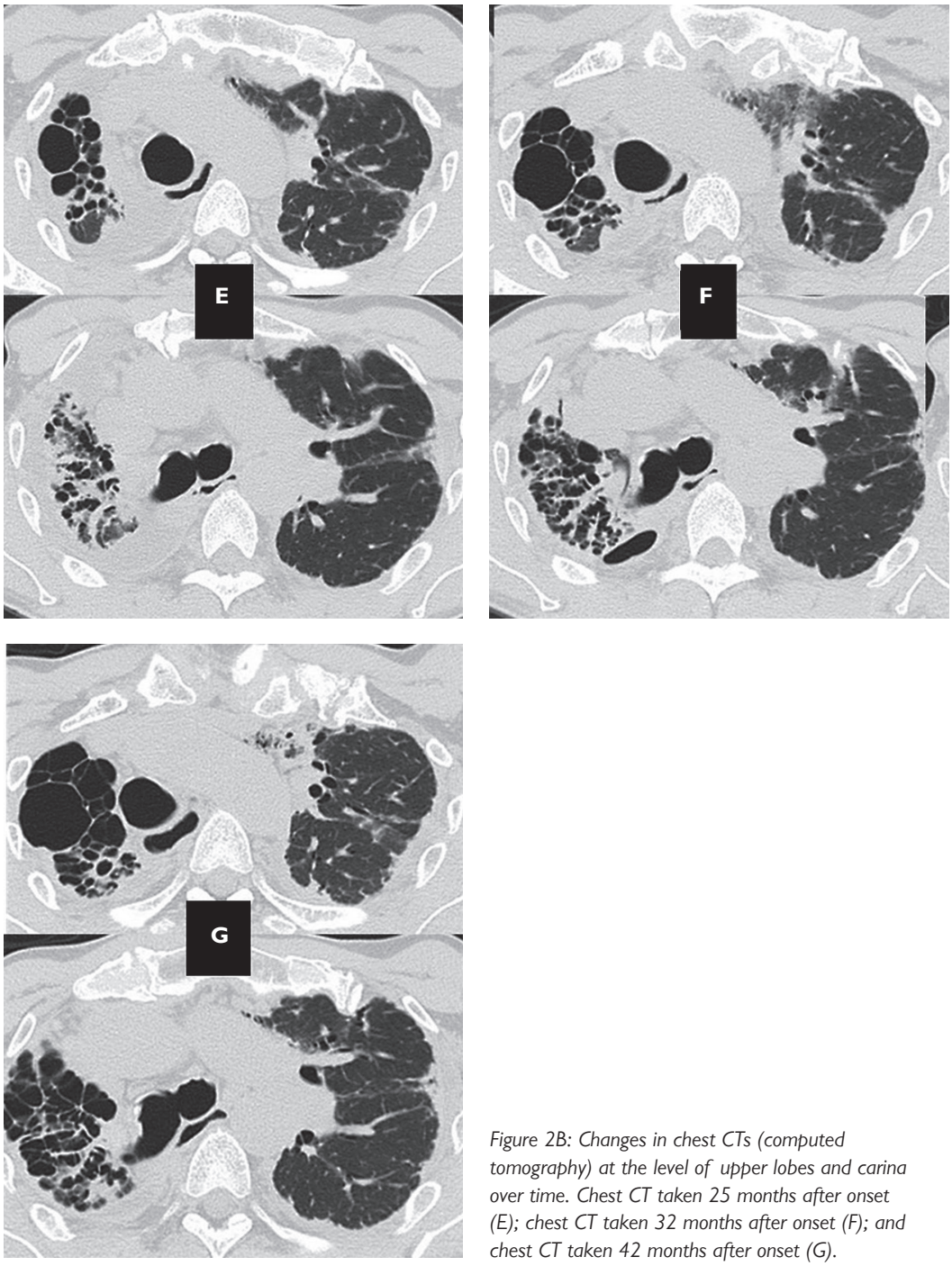


*Figure 2B: Changes in chest CTs (computed tomography) at the level of upper lobes and carina over time. Chest CT taken 25 months after onset (E); chest CT taken 32 months after onset (F); and chest CT taken 42 months after onset (G).*

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three years after onset, hypoxemic respiratory failure progressed, and home oxygen therapy was initiated. At this time, the patient's body weight was 48 kg, and although there was no pretibial edema, there was a clear decrease in muscle mass of lower extremities. The time course of chest X-rays and computed tomography (CT) scans are shown in Figures 1 and 2. Two years later, the patient developed a right pneumothorax, but recovered conservatively. His hypoxemic respiratory failure then progressed further, and he passed away due to respiratory failure 43 months after the onset of PPFE.

## Discussion

PPFE was included as one of the rare interstitial pneumonias in the International Classification of idiopathic interstitial pneumonias proposed in 2013 (Travis et al., 2013). It is a chronic, progressive pulmonary fibrosis, and its clinical and pathological condition are similar to idiopathic pulmonary fibrosis, but its histological characteristic is the proliferation of elastic fibers under the pleura of the bilateral upper lung fields (Kinoshita et al., 2017). PPFE is also a disease concept that overlaps with limited upper lobe pulmonary fibrosis (Amitani disease) reported in 1992 by Amitani et al. Most patients with PPFE develop bilateral apical pleural thickening and upper lobe fibrosis. Usual interstitial pneumonia, the most common type of pulmonary fibrosis, is characterized by prominent lesions in both lower lobes, traction bronchiectasis, thickening of broncho-vascular bands and honeycombing. However, in PPFE, lower lobe involvement is absent or mild, and these characteristic findings of idiopathic pulmonary fibrosis are usually not observed (Nakamura et al., 2021; Cottin et al., 2022).

The first noteworthy point in this case was that fibrotic change was prominent in right lung. Regarding the uneven distribution of fibrotic change, reports have pointed out the similarity between patients with unilateral upper lung PF (pulmonary fibrosis) and PPFE patients with prominent fibrosis in one lung (Sekine et al., 2017; Inafuku et al., 2022). In our patient's case, a progression was observed on images over time, from a time point when no fibrotic changes were evident. The progression of the fibrotic change over time in this patient was very similar to that observed in those with unilateral upper-PF (Sekine et al., 2017; Inafuku et al., 2022). The imaging course of this patient supported the notion that some PPFE patients had similar degrees of fibrosis on both sides and others had more prominent fibrotic change on one side.

The second point to be worthy of attention in our case was his medical history of surgery for renal cancer 64 months before the onset of PPFE. In previous reports by Sekine et al. (2017), the majority of patients with unilateral upper-PF had a medical history of thoracic surgery (Inafuku et al., 2022). Patients undergoing thoracic surgery included not only lung cancer patients but also patients with benign diseases, and this suggested that there was no direct association between unilateral upper-PF and malignant disease (Sekine et al., 2017; Inafuku et al., 2022). Our patient had no history of thoracic surgery but had undergone truncal surgery. Although we found no reports suggesting an association between abdominal surgery and unilateral upper PF, this medical history in our patient was intriguing.

The third notable point of the clinical course of this PPFE patient was the poor prognosis. The prognosis of PPFE remains poor at present, with the median survival time from diagnosis reported to be 3–8 years (Enomoto et al., 2017, Sekine et al., 2017; Ishii et al., 2018; Oda et al., 2021). No specific clinical factors have been found to be associated with a good or poor prognosis in this disease entity. In general, many patients with chronic respiratory disease have a poor prognosis due to weight loss or malnutrition, and these factors are also attracting attention in relation to the prognosis of interstitial lung diseases (Kinoshita et al., 2023; Suzuki et al., 2023). In our patient, the BMI was 19.9 at the time of his first visit to our hospital, and three years later, his BMI had decreased to 18.1. In relation to prognosis, in this patient, antifibrotic drug was initiated during the course of PPFE. It is unclear whether the course of the disease might have been different if this drug treatment had been initiated at the same time as the diagnosis of PPFE. Some studies have shown that PPFE is less responsive to antifibrotic drugs and has a poor prognosis (Sugino et al., 2021; Kinoshita et al., 2022). On the other hand, there are studies that show that antifibrotic drug treatment may slow the decline of lung function (Nasser et al., 2021; Cottin et al., 2022). As they were all small retrospective studies, further evaluation is required to determine whether the effectiveness of antifibrotic drugs in PPFE and other progressive pulmonary fibrosis is equivalent.

## Conclusion

We reported the course of a patient with pleuroparenchymal fibroelastosis. This patient had prominent right-sided lesions, suggesting similarity to unilateral upper pulmonary fibrosis. This patient had no history of thoracic surgery, but had undergone renal cancer resection, which was suspected to be related to the development of unilateral upper PF.

The prognosis for PPFE remains poor, as was the case in this patient. It is strongly expected that knowledge regarding the pathology and treatment of this disease will be accumulated, and thus the prognosis would be improved.

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# Thrombosis of the Princeps Pollicis Artery of the Thumb – Case Report of an Unusual Disease

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**Key words:** Thrombosis – Princeps pollicis artery – Ultrasonography – Doppler

**Abstract:** In the clinical evaluation of upper extremity embolism cases, the anamnesis should focus on identifying potential triggering risk factors. The physical examination may reveal sensory deficits, aiding in the determination of ischemia stages. Imaging diagnosis is crucial, with computed tomography (CT) angiography being the preferred examination due to its ability to provide detailed information about arterial anatomy across multiple planes and clear visualization of adjacent structures. Compared to magnetic resonance imaging angiography, CT angiography offers faster results with minimal distortion, despite the exposure to radiation and contrast use. Doppler ultrasonography is another valuable tool in suspected arterial thromboembolism cases, particularly in emergency settings. It offers advantages over CT angiography as it is non-invasive, cost-effective, and does not involve radiation or contrast administration. We present the case of a 68-year-old man who reported a nodule in the medial region of his right thumb for three months. Initially, he experienced significant local pain and limited movement, which gradually improved over time with the use of analgesic medication. Ultrasonography revealed thrombosis in the princeps pollicis artery, and the patient commenced treatment with acetylsalicylic acid. After four months, the patient reported a marked reduction in the nodule size along with pain improvement.

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## Introduction

Arterial occlusion in the upper limbs has different origins, such as arterial embolism, iatrogenic or traumatic arterial injury, native arterial thrombosis and arteritis, and most often has an acute clinical presentation. Native arterial thrombosis in the upper limbs can originate of atherosclerotic plaque, aneurysm thrombosis, arterial compression, thrombophilia or low blood flow state (Deguara et al., 2005; Bae et al., 2015).

The ulnar and radial arteries unite in the palm of the hand and form the deep palmar arch and the superficial palmar arch. The deep palmar arch is supplied by the radial artery and gives rise to princeps pollicis artery and the radial digital artery of the index finger. Princeps pollicis artery divides into two palmar digital arteries (Miletin et al., 2017). It's estimated that thrombosis in the upper limbs is six times rarer than thrombosis in the lower limbs (Chitte et al., 2003).

Herein we report the case of a 68-year-old patient who attended outpatient care complaining of pain in his right thumb.

## Case report

A 68-years-old man reports a nodule in the medial region of the thumb of his right hand for three months. Initially with marked local pain and movement limitation, evolving with improvement over time associated with the use of analgesic medication. He denies trauma at the beginning of the symptoms.

On physical examination, he presents a hardened nodulation that is slightly painful on palpation, on the medial portion of the thumb, without local hematoma and without changes in sensitivity or temperature in the thumb, with preserved strength and movement. Ultrasonography demonstrates thrombosis in princeps pollicis artery (Figure 1).

The patient started treatment with acetylsalicylic acid. After four months, the patient reports a marked reduction in the nodule with improvement in pain.

## Discussion

In the clinical evaluation of cases of embolism in the upper extremities, the anamnesis must be guided by the possible triggering risk factors. The physical examination may or may not indicate sensory deficits, determining the stages of ischemia (Rutherford, 2009). Laboratory markers aren't specific for the diagnosis of arterial embolism (Cooke and Wilson, 2010).

Computed tomography (CT) angiography is usually the choice exam, as it provides information about the arterial anatomy in multiple planes, in addition to presenting adjacent structures with good definition. CT angiography, in comparison with magnetic resonance imaging (MRI) angiography, is faster and have less distortion of the images, despite exposure to radiation and the use of contrast (Olin et al., 2004). The use of Doppler ultrasonography in cases of suspected arterial thromboembolism can also be fast in emergency scenarios and has advantages because it's non-invasive, cheaper, doesn't require contrast and is radiation-free (Crawford et al., 2016).

The diagnostic hypothesis for the condition presented by the patient was, initially, a nodule of etiology to be clarified. Initially, princeps pollicis artery thrombosis was not considered. Differential diagnoses, however, vary among artery occlusion, stenosing tenosynovitis, Bennett's fracture, anatomical variations in vascularization that facilitate the reduction of arterial supply to the hand, atheroma plaque in vessels that are the source of embolism and princeps pollicis artery pseudoaneurysm (Parks et al., 1978).

Ezomo et al. (2020) also reported a 61-year-old man referring that his right thumb turned blue the previous morning and had remained blue until the presentation

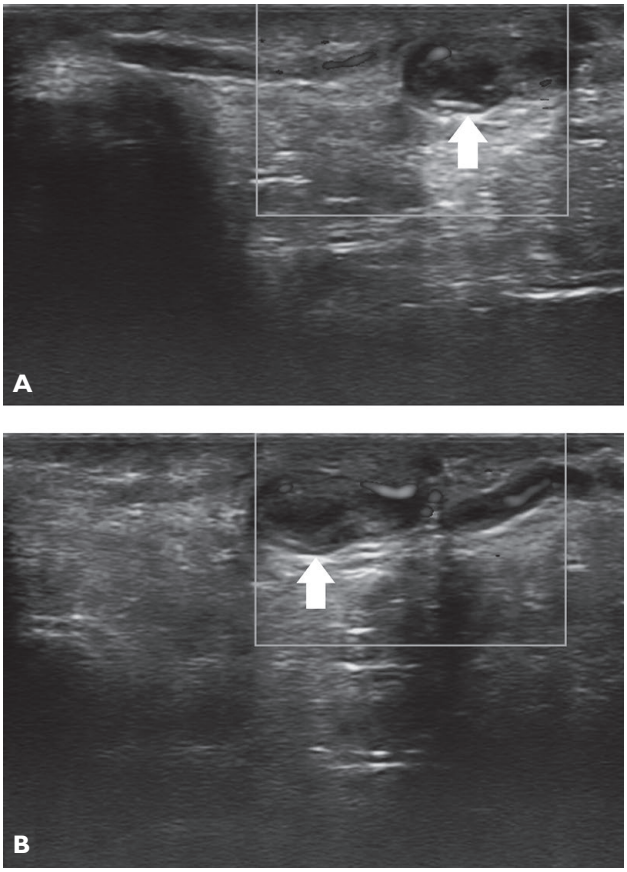


*Figure 1: Ultrasonography of the thumb demonstrating thrombosis of the princeps pollicis artery of the thumb (white arrow).*

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to the physician. The patient stated that his right thumb was cold. The princeps pollicis artery thrombosis was diagnosed by ultrasonography and, also, treated conservatively with clopidogrel in addition to acetylsalicylic acid. The patient experienced full resolution of thumb discoloration over the next few days and experienced no complications or recurrence at follow up visits (Ezomo et al., 2020).

The initial management of princeps pollicis artery thrombosis relies on systemic anticoagulation at diagnosis. The subsequent approach and the need for revascularization are guided by the individual's degree of severity, presence or absence of sensory and motor deficits, and condition of preservation of the flow. Other alternatives are transcatheter embolectomy and catheter-directed thrombolysis (Rutherford, 2009).

## Conclusion

The princeps pollicis artery is the main artery of the thumb, but its thrombosis is rare, having just case reports in the medical literature. Although the clinical presentation of the pathology is, in most cases, acute, the patient reported had preserved sensitivity, temperature and movement, in addition to the absence of limb ischemia. For this unusual form of occurrence, Doppler ultrasonography meets the diagnostic needs, without the necessity of the CT angiography or MRI.

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# Eccrine Hidrocystoma of Eyelid Masquerading as Epidermal Inclusion Cyst: A Rare Case Report with Review of Literature

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**Key words:** Eccrine hidrocystoma – Apocrine hidrocystoma – Eyelid swelling

**Abstract:** Eccrine hidrocystomas are rare, benign cystic lesions that usually affect the scalp, cheeks, and eyelids. They are thought to originate from the sweat glands. These lesions can be single or many in nature and frequently worsen in the summer from increased perspiration. They are caused by dilated ducts of eccrine sweat glands. Clinically, they seem like small, transparent cystic lesions that are painless, and they usually affect middle-aged or older people. However, a histological study is necessary to make a final diagnosis. We present a case of a 35-year-old woman who had an epidermal inclusion cyst first identified as a single, painless cystic growth on her right lower eyelid.

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## Introduction

Eccrine hidrocystomas are uncommon benign cystic lesions originating from the sweat glands that typically affect the eyelids and also occur on the face and scalp. Occurs mainly due to dilated ducts of eccrine sweat glands (Sarabi and Khachemoune, 2006; Kumar et al., 2021).

Hidrocystomas may occur singly or can present as multiple lesions (Singh et al., 2005). The lesions of eccrine hidrocytomas have a chronic course with periodic flares in summer months, associated with exacerbation in sweating (Alfadley et al., 2001).

The typical clinical presentation includes a painless, small-sized, translucent cystic lesion on the face, especially on the eyelid commonly affecting middle-aged or elderly persons. However, histopathological examination is needed to rule out other differential diagnosis.

We present a case of lower eyelid enlargement in a 35-year-old female diagnosed with eccrine hidrocystoma on histopathology.

## Case report

A 35-year-old female presented to the Department of Ophthalmology with a complaint of solitary, painless, 5×4 mm sized cystic swelling over her right lower eyelid for 2 months (Figure 1A). She has no other systemic complaints, no relevant family or personal history or any history of trauma. Based on clinical

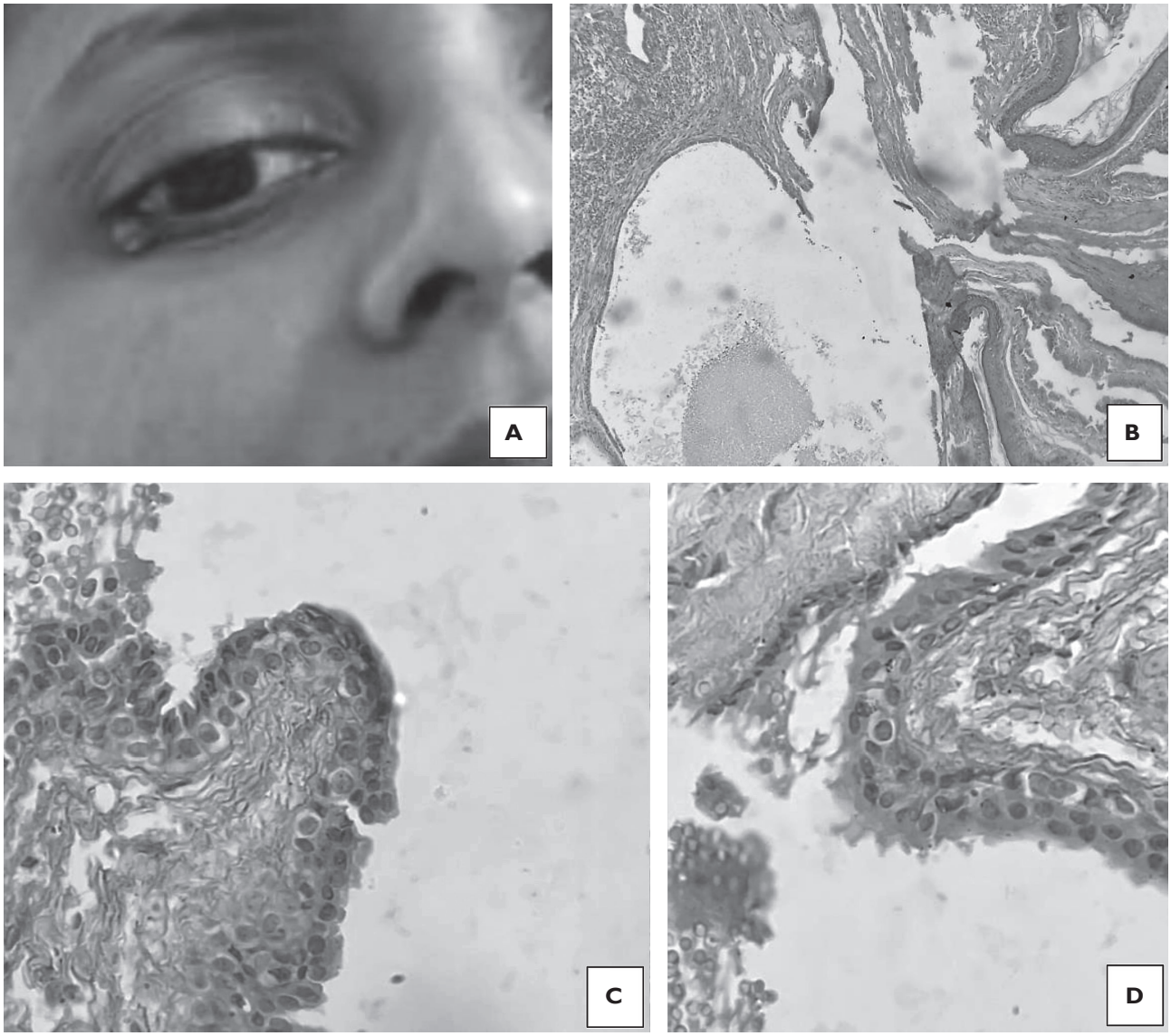


*Figure 1: (A) The photographic image shows a 5×4 mm sized cystic swelling over right lower eyelid. (B) Shows stratified squamous epithelium with presence of a cystic structure underneath it and filled with secretions (hematoxylin [H] and eosin [E] stain, 10×). (C and D) The cyst was lined by a double layer lining epithelium; outer flattened myoepithelial and inner cuboidal cells layer with eosinophilic cytoplasm (H and E stain, 40×).*

findings a diagnosis of epidermal inclusion cyst was made. Excision of the cyst was done and sent for histopathology. While excision straw coloured fluid came out from the cyst.

Histopathological examination of the tissue was done which revealed squamous lining epithelium, underneath which there is a cystic structure lined by double lining epithelium; the outer layer is lined by flattened myoepithelial cells, inner layer has cuboidal epithelial cells with eosinophilic cytoplasm and round to oval bland nuclei. There were no papillary projections or significant decapitation. Intraluminal secretions are also present in cysts. There is also the presence of chronic inflammatory cells comprised of lymphocytes in the surrounding stroma. These features favoured the diagnosis of eccrine hidrocystoma (Figure 1B–D).

## Discussion

Benign lesions of the eyelids are much more frequent than the malignant lesions. The differential diagnosis of eccrine hidrocystoma clinically includes follicular-derived cysts, epidermal inclusion cysts, haemangioma, lymphangioma, apocrine hidrocystoma, and nodular basal cell carcinoma in the early stages (Jaifi et al., 2024). Hence it is important to do histological analysis of all lesions removed from the palpebral region.

The eccrine hidrocystoma must be differentiated from its apocrine variant, which is considered its close differential diagnosis. The eccrine hidrocystoma usually does not involve the margin of eyelid, unlike apocrine hidrocystoma because the eccrine sweat glands are distributed throughout the eyelid skin and are not confined to the eyelid margin unlike the apocrine glands (Rodallec et al., 2006). However, histopathology is the only way to confirm the diagnosis and differentiate between these two entities.

On histopathological examination eccrine hidrocystoma consist of a cystic structure lined by a double-lining epithelium, with outer flattened myoepithelial layer and inner cuboidal cell layer with eosinophilic cytoplasm, but without papillary projections or characteristic decapitation secretions is present (Adenis et al., 1998).

Apocrine hidrocystoma also had a double layer lining epithelium. The outer layer consisting of myoepithelial cells and an inner layer composed of secretory columnar cells with eosinophilic cytoplasm and a characteristic apical decapitation secretions with papillary protrusions in lumen (Adenis et al., 1998; Armstrong et al., 1998).

Schöpf-Schulz-Passarge syndrome (SSPS), a rare kind of ectodermal dysplasia, is a collection of genetic illnesses characterised by developmental anomalies in two or more of the following structures: hair, teeth, nails, sweat glands, and other ectodermal-derived structures. SSPS is an autosomal recessive syndrome characterised by palmoplantar keratoderma, hypodontia, hypotrichosis, nail dystrophy, and numerous periocular and eyelid apocrine hidrocystomas (Schöpf et al., 1971; Hampton et al., 2005).

Hidrocystoma is generally treated surgically, especially in the case of single lesions that are large and cause functional damage. Eccrine hidrocystomas, present no risk of malignant transformation, but a risk of recurrence is observed in 2.3% of cases when there is incomplete excision or cyst rupture while excision with residual cyst wall (Schöpf et al., 1971; Adenis et al., 1998).

## Conclusion

Eccrine and apocrine hidrocystomas are rare cystic benign lesions of the sweat glands occurring on the face, especially over the eyelid. Histopathological examination plays an important role as the entity can mimic other lesions of the eyelids which also include malignant lesions. Chances of recurrence are comparatively high if the object is not completely removed on excision.

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# Pronator Teres Syndrome – Case Report with Imaging Tests Diagnosis

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**Key words:** Median nerve – Ultrasonography – Diagnosis

**Abstract:** Pronator teres syndrome is characterized by compression of the median nerve, leading to dysfunction of the affected limb. Median nerve entrapment causes paresthesia, changes in sensitivity, and loss of strength in the fingers, in addition to causing loss of hand dexterity. The diagnosis of pronator teres syndrome is complicated, due to its similarity with other neuropathies of the median nerve. So, it is important to emphasize the need for a physical examination together with imaging tests, especially ultrasound, for its correct diagnosis. We report the case of a 28-year-old woman who complained of tingling for ten years in the proximal third of the left forearm at rest that worsens on exertion and weakness if not moving. On physical examination, she has no limitation of movement but refers to a feeling of weakness and numbness in his forearm. Ultrasonography demonstrates compression of the median nerve between the ulnar and humeral heads of the pronator teres muscle, a finding confirmed by magnetic resonance imaging and electroneuromyography. The patient was treated with physiotherapy presenting improvement of symptoms after 45 days.

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## Introduction

Anatomically, the median nerve originates from the brachial plexus, crossing several structures (Soubeyrand et al., 2020), such as the ligament of Strutters, the two bellies of the pronator teres muscle, the lacertus fibrosus, the arcade of the superficial flexor digitorum, the origin of the anterior interosseous nerve and the vascular arches (Dubois de Mont-Marin et al., 2021). Thus, the median nerve is susceptible to compression under any alteration of some of these structures (Binder et al., 2021).

Pronator teres syndrome is a rare condition (Moura and Agarwal, 2020) and its etiology is the hypertrophy of the pronator teres muscle or schwannoma in the pronator tunnel (Binder et al., 2021), caused by repetitive movements of supination and pronation, overload or trauma with associated hematoma or deformity (Babaei-Ghazani et al., 2018; Chang et al., 2022). This entrapment commonly occurs between the humeral heads and ulnar of the pronator teres muscle (Binder et al., 2021). However, this entrapment can occur due to other proximal anatomical structures in the upper forearm (Moura and Agarwal, 2020).

For the diagnosis, it is necessary to investigate the signs and symptoms – sensory alteration, pain, paresthesia, and alteration in finger movement – together with ultrasound, due to the symptomatologic similarity with carpal tunnel syndrome (Babaei-Ghazani et al., 2018).

Herein, we report the case of a 28-year-old male patient reporting tingling in the proximal third of the left forearm at rest that worsens on exertion and weakness in the hand.

## Case report

A 28-year-old man complaining of tingling for ten years in the proximal third of the left forearm at rest that worsens on exertion and weakness if not moving. He denies previous surgeries and traumas. Denies diseases under treatment. On physical examination, he has no limitation of movement but refers to a feeling of weakness and numbness in his forearm. The rest of the physical exam is normal.

Ultrasonography demonstrates compression of the median nerve between the ulnar and humeral heads of the pronator teres muscle (Figure 1), a finding confirmed by magnetic resonance imaging (Figure 2). The electromyography showed prolonged sensory latency in the distribution of the median nerve corresponding to the pronator teres region.

The patient started physiotherapy, without medications, with improvement of symptoms after 45 days, not showing numbness at rest, referring it only to great efforts. Thus, the patient refused surgical treatment.

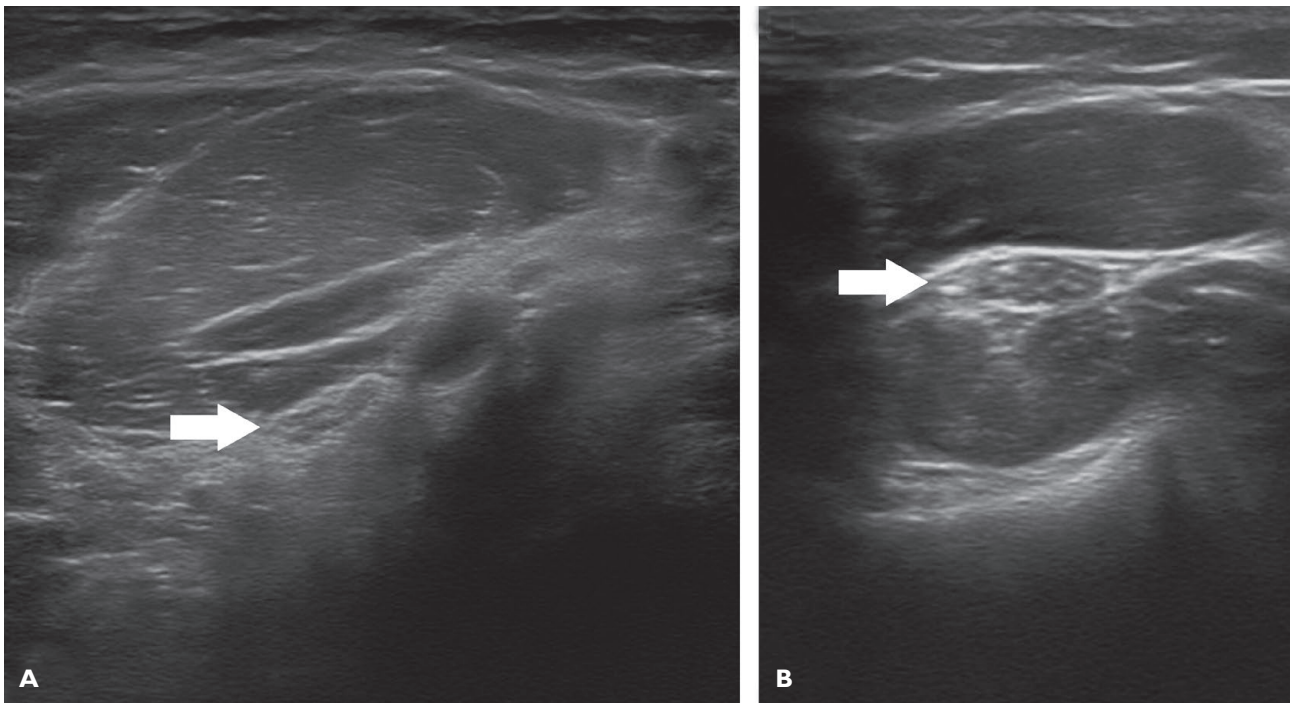


*Figure 1: A) Ultrasonography of the left forearm demonstrating flattening of the median nerve between the muscle bellies of the humeral and ulnar heads of the pronator teres (white arrow). B) Ultrasonography of the left forearm at the level of the radial head, demonstrating a normal-looking median nerve (white arrow).*

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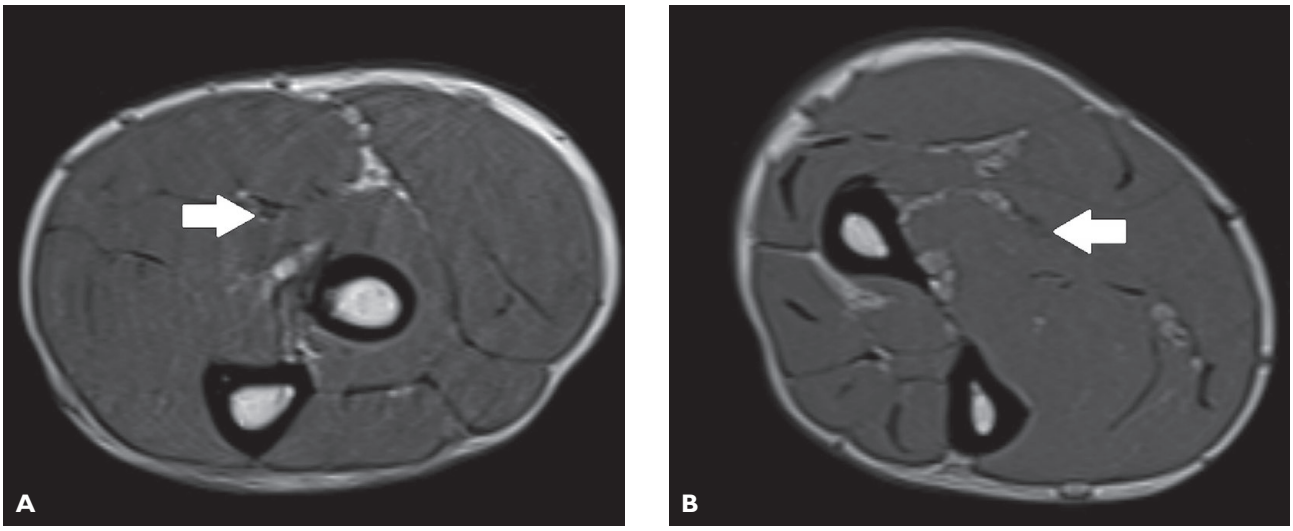


*Figure 2: A) Magnetic resonance imaging of the right forearm demonstrating the usual-looking median nerve (white arrow). B) Magnetic resonance imaging of the left forearm demonstrating flattening of the median nerve between the muscle bellies of the humeral and ulnar heads of the pronator teres (white arrow).*

## Discussion

The peripheral nerves are easily translocated in the intramuscular compartments (Chang et al., 2022). However, in pronator teres syndrome the median nerve is compressed by adjacent structures, either by the pronator teres muscle, Strutters ligament, bicipital aponeurosis, or the fibrous border of the flexor digitorum muscle (Babaei-Ghazani et al., 2018). So, symptoms are generally caused by the involvement of the flexor pollicis longus muscles and the radial half of the flexor digitorum profundus muscles (Chang et al., 2022).

Clinically, it is described as a median neuropathy of the proximal forearm (Soubeyrand et al., 2020), with symptoms of pain in this region, sensory alteration of the first to third radial digits, paresthesia, weakness of the intrinsic muscles of the hand, loss of strength in pinching, loss of agility with the hand (Babaei-Ghazani et al., 2018), dysesthesia or paralysis in the innervation zone of the median nerve (Binder et al., 2021), as demonstrated in the case report by Moura and Agarwal (2020).

The isolated pronator teres syndrome is a rare disease, which has a complicated differential diagnosis from other neuropathies of the median nerve, such as the usual carpal tunnel syndrome, double crush syndrome, and anterior interosseous nerve syndrome (Moura and Agarwal, 2020). The occurrence of often from the association between carpal tunnel syndrome and pronator teres syndrome (Binder et al., 2021).

In addition to the symptoms, tests can be performed in the physical examination, such as resisted forearm pronation test, middle finger test, and resisted flexion-supination test (Binder et al., 2021) and imaging exams, with emphasis on ultrasound or magnetic resonance (Créteur et al., 2017). Electromyography is also useful for establishing the diagnosis (Créteur et al., 2017), but abnormal electrodiagnostic findings are observed in only 10% of pronator teres syndrome cases (Afshar, 2015).

It is important to emphasize that ultrasound is essential for the diagnosis of peripheral nerve compression, above all, showing a change in the cross-sectional area of the nerve. In technical terms, the reference value of the median nerve is 4.9–12.9 mm, and the difference in the area of the median nerve between the upper limbs of up to 3.0 $mm^2$ at the level of the pronator teres muscle (Chang et al., 2022). Créteur et al. (2017) suggest the evaluation of the upper limbs with four cross-sectional images complemented by dynamic tests. Ultrasonography allows standard and dynamic evaluations of the median nerve, in addition to post-traumatic lesions of the pronator teres muscle and local tumours.

Finally, the treatment is based on physiotherapy and surgery, so that the surgical procedure may be elective depending on the state of the syndrome in the patient (Binder et al., 2021).

## Conclusion

Pronator teres syndrome is a rare disease that is difficult to diagnose, and generally hinders the dexterity of hand movements. For its investigation,

it is necessary to correlate the physical and imaging exams and present the surgical procedure as the main treatment. But, as demonstrated in the case reported, physiotherapy can improve the symptoms, without the need for surgery.

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# Two Cases of Sternalis Muscle in Humans: Clinical Considerations

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**Key words:** Sternalis muscle – Sternalis – Pectoral region

**Abstract:** The sternalis is an occasional muscle of the pectoral region with a reported incidence of around 7.8%. Higher rates of occurrences have been noted in females and in Asians. The muscle becomes clinically relevant as the muscle can be mis-interpreted as a pathological mass. Routine dissection was performed in a 68-year-old male and a 65-year-old female donated cadavers in the pectoral region. Gross anatomical features were meticulously noted, photographed and measurements were recorded with digital Vernier callipers. In the male cadaver, a long, slender, flat bi-tendinous sternalis was found, and in the female cadaver, tripartite sternalis possessing three distinct bellies was observed. Both the sternalis was right sided and were supplied by 2$^{nd}$, 3$^{rd}$ and 4$^{th}$ intercostal nerves. Single bellied and tripartite sternalis are rare and they need to be recognized in the diagnostic images and during surgeries to avoid confusion.

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## Introduction

The rectus sternalis or simply sternalis is an occasional muscle found in the pectoral region of humans lying superficial to the pectoralis major. The sternalis muscle often has continuity with the rectus abdominis muscle and the external oblique aponeurosis. Superiorly the muscle is attached with the sternal origin of the sternocleidomastoid muscle or to the pectoral fascia. The earliest description of the sternalis muscle was by Cabrolio, a French surgeon in 1604. Detailed morphology of the muscle was elaborated by Du Puy in 1726 (Raikos et al., 2011; Snosek et al., 2014). The muscle has been reported across all races and known by other names such as the sternalis, the rectus thoracis, rectus sterni, superficial rectus abdominis, episternalis, parasternalis, presternalis, sternalis brutorum etc. The reported incidence is approximately 7.8% with higher rates of occurrences in females and in Asians (Raikos et al., 2011; Snosek et al., 2014). Unilateral sternalis is more common than bilateral. Pectoral or intercostal or both these nerves often innervate the muscle. It was Cunningham in 1884, who systematically traced the nerves supplying the sternalis back to the medial pectoral nerve. The muscle becomes clinically relevant as the muscle can be wrongly interpreted as a pathological mass (Snosek et al., 2014). Two cases of right sided sternalis muscle – one single bellied and one tripartite, both supplied by 2nd, 3rd and 4th intercostal nerves in a male and a female cadaver are presented here.

## Case report

During routine dissection of the pectoral regions of a male cadaver aged 68-years and a female cadaver aged 65-years, right sided sternalis muscles were observed. Gross anatomical features were meticulously noted, photographed and measurements were recorded with digital Vernier callipers.

### Case 1

A long flat bi-tendinous muscle was observed in the right parasternal region of the male cadaver. The muscle was identified as the sternalis and it belonged to the type I [1] as per the classification system of Jelev et al. (2001). Superior tendinous potion originated from the pectoral fascia overlying the manubrium of the sternum followed by a flat fusiform muscle belly and eventually inserted via a thin flat tendon merging with the pectoral fascia, perimysium

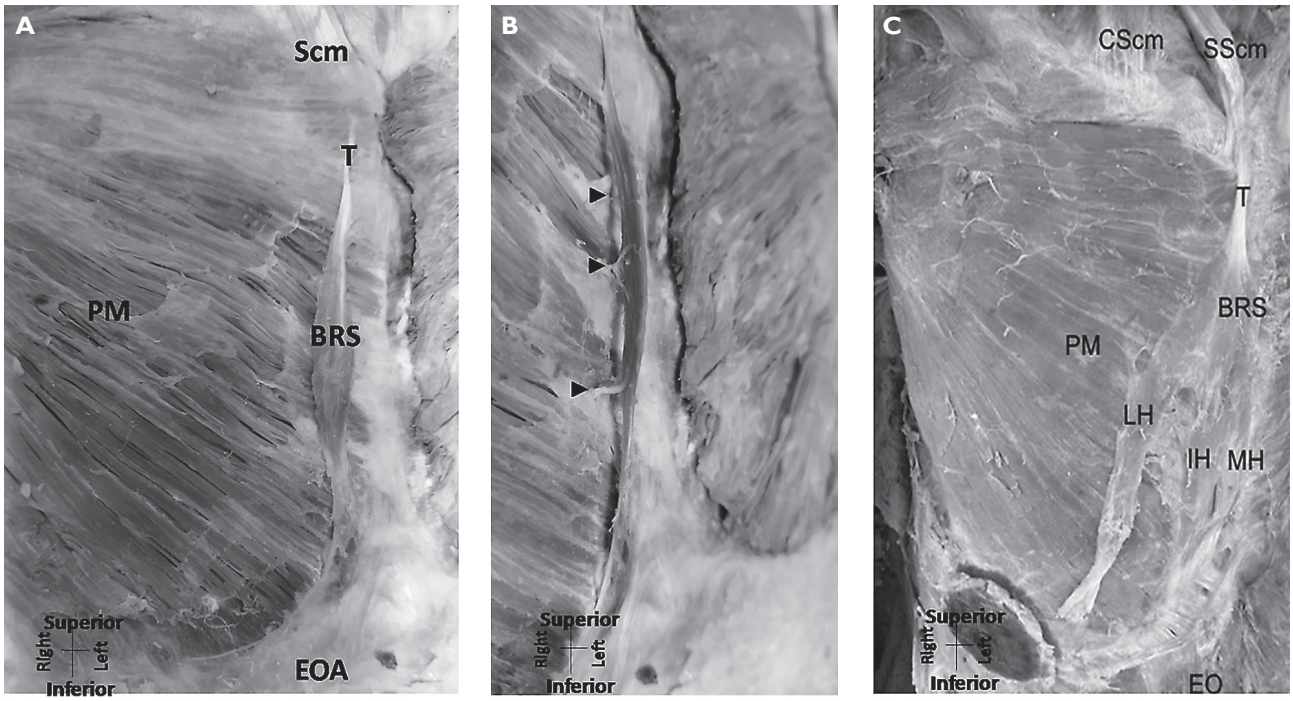


*Figure 1: Photograph of simple and mixed sternalis muscle in two cases. A) Right sided single bellied sternalis muscle in case 1. B) Sternalis muscle in case 1 seen innervated by intercostal nerves. C) The mixed type of right sided multiple bellied sternalis muscle in case 2. Scm – sternocleidomastoid; CScm – clavicular head of sternocleidomastoid; SScm – sternal head of sternocleidomastoid; T – tendon of sternalis; BRS – belly of sternalis; LH – lateral head; IH – intermediate head; MH – medial head; EO – external oblique muscle; EOA – external oblique muscle aponeurosis; PM – pectoralis major muscle; black arrowheads indicate the intercostal nerves supplying the sternalis muscle.*

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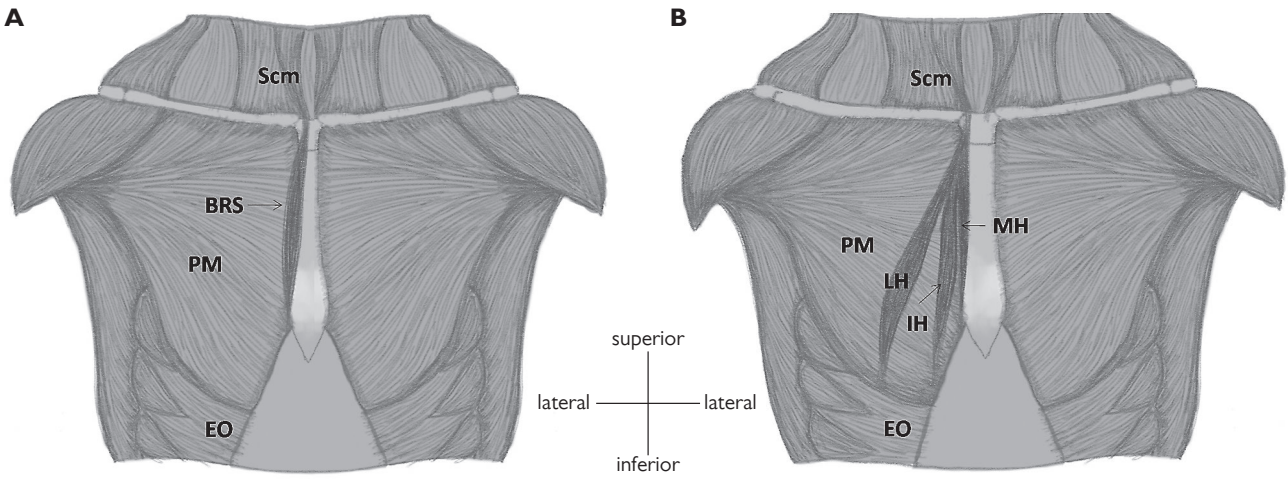


*Figure 2: Morphology and location of two variants of unilateral sternalis muscle observed in this report. Scm – sternocleidomastoid; BRS – belly of sternalis; PM – pectoralis major muscle; EO – external oblique muscle; LH – lateral head; IH – intermediate head; MH – medial head.*

covering the external oblique and the rectus abdominis muscle (Figure 1A and B). Muscle fibres were directed cranio-caudally. The superior tendon, muscle belly and the inferior tendon measured 3.5×0.47 cm, 4.6×2.3 cm and 3.1×0.54 cm respectively in the greatest dimensions. The muscle was innervated by the branches of 2nd, 3rd and 4th intercostal nerves.

**Case 2**

In the female cadaver, a right sided sternalis with unique tripartite configuration was observed. It had three distinct bellies originating from a common tendinous origin at the sternal head of the sternocleidomastoid muscle. The bellies were diverging in nature and were arranged as follows: a medial head directed towards the xiphoid process, an intermediate head coursing vertically downward towards the external oblique aponeurosis and a lateral head directed inferolaterally towards the right nipple (Figure 1C). Each of these heads had a well-defined muscle belly and was positioned between the superficial fascia and the pectoral fascia. The dimensions of the medial, intermediate and the lateral head were 1.02×4.12 cm, 1.87×4.62 cm and 1.17×4.92 cm respectively. All the bellies of this muscle were supplied by the branches of 2nd, 3rd and 4th intercostal nerves. Illustrative sketch of the two variants of sternalis has been provided for easy understanding (Figure 2A and B).

## Discussion

The prevalence of unilateral and bilateral sternalis is 67% and 33%, and among unilateral cases more instances have been noticed on the right side. Since its first description in 1604, the various morphology of the muscle with different neurovascular supply has been reported. Initially sternalis variants were classified into type I and type II categories by Jelev et al. (2001), which was expanded by Ge et al. (2014) and re-classified into three major types and three subtypes. Later Snosek et al. (2014) modified the classification system, but, for categorization of the present two cases we are referring to the Jelev et al. (2001) and the Ge et al. (2014) classification systems. The sternalis in the male cadaver can be categorized into type I [1] variety according to the Jelev et al. (2001) system and type I A as per the Ge et al. (2014) system. The second case of tripartite sternalis in the female cadaver partially resembles type I [2] variety according to the Jelev et al. (2001) proposed classification system and type III A as per the Ge et al. (2014) proposed system. The tripartite configuration observed in the female cadaver does not exactly match to any of the previously described variety. However, it resembles partially to a recently described new variant of sternalis by Dudgeon et al. (2017) which they named as the triple subtype.

Sternalis muscle is often found associated with other coexisting anomalies. Recently bilateral sternalis muscle has been reported with co-occurrence of asymmetrical bilateral extensor carpi radialis intermedius and with aberrant pectoral muscle (Shekhawat et al., 2023; Boadum and Lu, 2024). Regarding origin of sternalis, Cunningham hypothesized that the muscle is a displaced portion of the pectoralis major disposed anterior to it due to rotation. Still some others believed that the sternalis had formed from the rectus sheath. On the other hand, many authorities consider that the sternalis could be a derivative of panniculus carnosus of lower

**Table 1: Mini literature review about the recently reported normal and symptomatic sternalis variants and their notable features**

| Authors and the population studied | Sample size and study type | Notable findings |
|---|---|---|
| Demirpolat et al. (2010); Turkish | 52,930, retrospective radiologic study | Sternalis was detected in 10 females out of the 52,930 examined mammograms which were identified as a small, asymmetric opacity at the medial posterior edge of the breast. |
| Mehta et al. (2010); Indian | 88, cadaveric study | 1 in 88 cadavers revealed unilateral sternalis (incidence = 1.14%). |
| Raikos et al. (2011); European | 45, cadaveric study | 1 in 45 revealed unilateral left sided sternalis (incidence = 2.2%). |
| Shiotani et al. (2012); Japanese | 948, retrospective radiologic study | Sternalis was more frequent in females (13.0%) compared to males (8.4%) with an overall incidence of 10.5%. |
| Anjamrooz (2013); Iranian | cadaveric case | Described biceps sternalis. |
| Ge et al. (2014); Chinese | 6,000, retrospective radiologic study | Sternalis was detected in 347 out of 6,000 (incidence = 5.8%). |
| Gruber et al. (2016); European | clinical case | Symptomatic sternalis in a 51-year-old female. |
| Dudgeon et al. (2017); American | clinical case | Bilateral sternalis in a 96-year-old female. |
| Davimes et al. (2018); African | cadaveric study | Incidence of unilateral sternalis 2.25%. |
| Al-Alami and Altamimi (2020); Jordanian | 1,709, retrospective radiologic study | Revealed 5.9% incidence of sternalis. |
| Shekhawat et al. (2023); Indian | cadaveric case | Bilateral sternalis on the right side. |
| Boadum and Lu (2024); American | cadaveric case | Bilateral sternalis was observed. |

mammals. Developmental origin could be either from the hypaxial myotomes or dermomyotomes (Paraskevas and Raikos, 2010). A mini literature review about the sternalis muscle reported during the last 15 years with their notable features is presented in Table 1. The nerve supply whether from the brachial plexus or from the intercostal nerve is still debated. In our cases the 2$^{nd}$, 3$^{rd}$ and 4$^{th}$ intercostal nerves supplied both the muscles.

Sternalis, especially unilateral, may mimic a mass lesion in imaging studies, more commonly a breast lump and can confuse clinicians. Pain due to sternalis has also been reported in a few cases which could be due to compression of nerve branches while coursing through the muscle. Gruber et al. (2016) described an interesting case of clinically symptomatic sternalis muscle in a female patient who presented with pain upon pressure. The sternalis muscle has the potential for utility as an autograft. As the sternalis is an occasional muscle, clinicians may not be aware about the muscle. A recent survey examined the awareness about the sternalis muscle among clinicians in Jordan. Surprisingly, very small fraction of the cohort could recognise the muscle from given anatomy dissection images and computed tomography images (Al-Alami and Altamimi, 2020). Hence it is pertinent to re-introduce this rare muscle once again, which is in fact already known in medical literature for over the last 400 years.

## Conclusion

Considering the fact that the pectoral region is being subjected to imaging studies and surgical procedures, it is possible that the aberrant muscle can be mis-interpreted for pathological mass. Thus, the knowledge about this aberrant muscle is very relevant for clinicians. The tripartite sternalis with single head of origin in the female cadaver of this report does not exactly resemble any known variety. Hence, we recommend amendment and inclusion of a separate category in the existing classification system of sternalis muscle.

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     Teich, N. (1984) Taxonomy of retroviruses. In: *RNA Tumor Viruses,* eds. Weiss, R., Teich, N., Varmus, H., Coffin, J., pp. 25–207, Cold Spring Harbor Laboratory, Cold Spring Harbor, New York.
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