



**Fig. 1.** Anteroposterior X-rays of the left (A) and right carpus (B) showing bilateral Madelung's deformity.

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## Atypical Cogan's Syndrome Associated With Sudden Deafness and Glucocorticoid Response\*



### Síndrome de Cogan atípico asociado a sordera brusca y respuesta a los glucocorticoides

Dear Editor,

Cogan's syndrome (CS), described by David G. Cogan in 1945,<sup>1</sup> is a disease of unknown etiology, but probably has an autoimmune basis. It is characterized by the association of non-syphilitic interstitial keratitis and audiovestibular symptoms (similar to Ménière's syndrome), occurring in a period of less than 2 years. It occurs predominantly in young adult Caucasians. There is no predominance of gender.<sup>2</sup> When the eye or audiovestibular manifestations are different from what is seen, or the interval between them is more than 2 years, it is called atypic CS.<sup>3</sup> It may be associated with other systemic manifestations such as aortitis and vasculitis.<sup>4</sup> A patient with atypical CS is presented.

The patient is a 44-year-old male, hypertensive, with axial psoriatic arthritis since age 20 and anterior nongranulomatous

uveitis developed in the last 4 years. He attended consultation derived from Otorrhinolaryngology for bilateral Ménière's syndrome resistant to standard therapy, of 3 years of evolution. Audiometry revealed cophosis stood in the left ear and moderate hearing loss in the right ear (RE) at 60 dB (the patient wore a hearing aid). Physical examination showed psoriasis and redness in the extremities of both ears, which were rubbery to the touch. No peripheral arthritis was found but he had axial involvement (BASMI: 6). No other data were found upon examination. Laboratory tests showed: ESR of 49 mm in the 1st hour and CRP 25 mg/L. CBC, serum biochemistry and urinalysis were normal. Autoimmunity: rheumatoid factor, antineutrophil cytoplasmic antibodies, antinuclear antibodies and anti-CCP antibody were all negative. HLA B27 antigen was positive. C3 and C4 were normal. Radiographs of the chest, hands and feet were normal. The sacroiliac X-rays showed stage IV sacroiliitis. Electrocardiogram, echocardiography and computed tomography with intravenous contrast were normal. Atypical CS was diagnosed associated with relapsing polychondritis (no biopsy was performed) and psoriatic arthritis. Methotrexate (MTX) was started orally at doses of 12.5 mg/week. At 2 months he presented sudden hearing loss in the RE compared to baseline. The bilateral otoscopy was normal. Audiometry revealed a serious 60 dB drop in treble in the RE conversational frequencies. ESR and CRP were 86 mm and 45.30 mg/L, respectively. Treatment was initiated with boluses of methylprednisolone 1000 mg every 24 h for 3 days, showing a good response, recovering

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RE hearing to baseline (determined through a new audiometry) and normalization of acute phase reactants. The patient was discharged with prednisone 30 mg/day and tapered the dosage of MTX to 12.5 mg/week, remaining asymptomatic after 6 months. Since the first description of CS, more than 220 cases have been described, 92 of them appearing atypically.<sup>5</sup> Unlike typical CS, the atypical variety is most commonly associated with systemic<sup>6</sup> manifestations and other autoimmune diseases, such as sarcoidosis, rheumatoid arthritis, relapsing polychondritis, juvenile idiopathic arthritis, Sjögren's syndrome and inflammatory bowel disease, among others.<sup>7</sup> Our case may raise doubts about the diagnosis, given the coexistence of several autoimmune<sup>8,9</sup> diseases. Psoriatic arthropathy could justify that the patient presented uveitis. Relapsing polychondritis can also present with hearing loss and vertigo, although generally it is a conductive hearing loss and vestibular dysfunction is not as similar to Meniere's. In this patient, the vestibular episodes were intense, with prolonged and bilateral sensorineural hearing loss, preceded by ocular involvement in less than a two year interval, and in the absence of specific complementary data, made us opt for the diagnosis of atypical CS, fulfilling the criteria established by Haynes et al.,<sup>3</sup> with 2 associated autoimmune disorders (psoriatic arthritis and relapsing polychondritis) and showing a good response to corticosteroid and immunosuppressive therapy, something relevant given the poor prognosis of deafness.

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## Usefulness of the Ankle-brachial Index as a Survey Method for Subclinical Vascular Disease in Patients With Rheumatoid Arthritis<sup>☆</sup>



### Utilidad del índice tobillo-brazo como método de cribado de enfermedad vascular subclínica en pacientes con artritis reumatoide

To the Editor,

Rheumatoid arthritis (RA) is a systemic inflammatory disease, with a chronic and variable evolution, characterized by persistent and symmetrical synovitis of the peripheral joints. In recent years its natural history has changed thanks to advances in treatment, so comorbidities have become more important; in fact, increased mortality compared to the general population is primarily a result of diseases of cardiovascular origin,<sup>1</sup> with rates up to 50% or higher.

In RA underlying atherosclerotic disease is increased<sup>2</sup> secondary to chronic inflammation, which involves activation of T lymphocytes and macrophages, production of proinflammatory cytokines<sup>3</sup> (gamma interferon, tumor necrosis factor, IL-1 and IL-6). It is potentiated due to classic cardiovascular risk factors (CVRF), including the metabolic syndrome, which is more prevalent probably due to less physical activity because of joint pain and moreover, dyslipidemia follows a more atherogenic<sup>4</sup> pattern.

With all these data, we conclude that the RA is a situation with a high CVR, where cardiovascular morbidity is related to the disease

activity, so its control could reduce the risk.<sup>5</sup> Therefore, this study proposes to detect subclinical CVD by measuring the ankle-brachial index (ABI).

We performed a descriptive cross-sectional study on 60 RA patients with no history of CVD, at the University Hospital of La Princesa, Madrid, selected consecutively in the rheumatology clinic during the 6 months when the study was carried out. Sociodemographic variables, analytical data, classic CVRF, duration of RA and immunomodulatory treatment were collected. ABI was defined as abnormal if less than 0.9.<sup>6</sup>

Of the 60 patients enrolled, 3 were men (5%) and 57 women (95%) with a mean age ± standard deviation of 53.75 years (53.75 ± 15.38, range 29–87). 38 had mild RA (63.3%), while 22 (36.7%) had important deformities. The time of disease progression was 9.14 years (9.14 ± 6.505, range 0.6–40), 58 patients (96.7%) were under immunomodulatory therapy, mostly with methotrexate (75%). The result of the ABI was similar in both lower limbs: 1.074 (1.074 ± 0.082, range 0.88–1.28) on the right and 1.077 (1.077 ± 0.088, range 0.92–1.27) on the left, with no significant differences between them. Only one patient (1.7%) had an abnormal ABI: a woman of 87 years, with hypertension, and RA for 12 years and using corticosteroids during virtually all this time; the ABI on the other extremity was 0.92. Fig. 1 shows the ABI results.

In our sample, there is an overrepresentation of women (19:1) with respect to other RA populations (3:1). However, the profile of CVR did not differ regarding the Spanish general population.<sup>7</sup> Only one pathological case was detected, much lower than other studies,<sup>8,9</sup> with rates of 20%–25%, although the frequency cutoff point considered as pathological was 1, rather than the value of 0.9 currently accepted. However, in another publication with the same cutoff,<sup>10</sup> the prevalence was 10%, although in their sample the mean age, duration of RA and, above all, the prevalence of cardiovascular

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