Diffuse idiopathic skeletal hyperostosis: time for a change.

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*The Journal of Rheumatology* is a monthly international serial edited by Earl D. Silverman featuring research articles on clinical subjects from scientists working in rheumatology and related fields.
Diffuse idiopathic skeletal hyperostosis (DISH) is a condition characterized by calcification and ossification of soft tissues, mainly ligaments and entheses. This condition was described by Forestier and Rotes-Querol more than 50 years ago, and was termed senile ankylosing hyperostosis. The axial skeleton is often involved, particularly the thoracic spine, but involvement of peripheral joints led researchers to use the name DISH. The main target of the disease process is within the entheses, an organ rich in collagen fibers, fibroblasts and other mesenchymal cells, fibrocartilage, and calcified matrix that penetrate the bone cortex at its attachment. Currently, the diagnosis of DISH is based upon classification criteria that require the presence of flowing osteophytes involving the anterolateral aspect of the thoracic spine. The lower thoracic spinal segment is usually the first to be involved, with subsequent extension into the upper thoracic segments and the lumbar spine. In the absence of validated diagnostic criteria, 3 sets of classification criteria are currently in use for the diagnosis of this condition. The classification criteria set by Resnick and Niwayama requires involvement of at least 4 contiguous vertebrae of the thoracic spine, preservation of the intervertebral disc space, and absence of apophyseal joints or sacroiliac inflammatory changes. Bridges connecting 2 vertebral bodies in at least 2 sites of the thoracic spine have also been suggested by Julkunen, et al to be characteristic for DISH. None of these sets of criteria took into consideration any of the peripheral manifestations of the condition. However, another set of criteria, defined by Utsinger as probable DISH, lowered the threshold for spinal involvement to 3 contiguous vertebral bodies, but added the presence of peripheral enthesopathies to the diagnostic measures. Despite the predilection to the thoracic spine, the peripheral joints are often affected by DISH. Enthesopathies with subsequent new bone formation, and stiffening of peripheral joints, generate features that distinguish them from primary osteoarthritis (OA). These include: a more frequent involvement of joints that are not usually affected in OA, such as metacarpophalangeal joints, elbows, and shoulders, and a more severe hypertrophic disease. Calcification and/or ossification of ligaments and entheses affecting the peripheral joints such as peripatellar, cruciate ligament insertion, and pericapsular osseous enthesopathies have all been described. Enthesal ossification of the heel, ribs, and pelvis are common findings in DISH and may become symptomatic, exhibiting pain in the affected region.

Of particular interest is the predictive value for the presence of DISH that was noted for ossification of the ilio-lumbar and sacrotuberous ligaments, and with bony overgrowth of the inferior acetabular rim. These features, together with the predilection to the thoracic spine, the preservation of the intervertebral disc height, a different prevalence and sex distribution, more hypertrophic bony changes of the involved joints, and involvement of joints usually not affected by OA, distinguish it from primary OA. Isolated involvement of the cervical spine has also been described. The various sites and aspects of peripheral involvement noted above and the involvement of spinal segments other than the thoracic spine are usually not taken into account for the diagnosis of DISH. It was suggested that a tentative diagnosis of DISH be made, on the basis of symmetrical and peripheral characteristic enthesopathies, even in the absence of spinal involvement.

In common practice, radiographs are ordered for the affected areas, while thoracic spine radiographs are seldom ordered unless the patient is symptomatic, or is highly suspected of having DISH based on the other manifestations of this condition. What would be, then, the diagnosis of an elderly, overweight patient with groin pain and a large ossification of the hip capsular enthesis without T-spine enthesal involvement? Recognition of DISH is important in several aspects. It can explain some clinical, otherwise unclear rheumatologic manifestations, and can avoid or change the attitude toward presence of future complications attributable to DISH such as dysphagia, unstable spinal fractures, spinal stenosis, postsurgical heterotropic ossifications, difficult intubation, difficult gastroscopy, aspiration pneumonia, myelopathy, and others. Diagnosing DISH may also expose some underlying correctable conditions such as dyslipidemia, hyperinsulinemia, hyperuricemia, hypertension, and others. Despite improvement in our understanding of DISH and its associated conditions, specific therapeutic interventions are not yet available, and correction of the
associated metabolic derangements is recommended. It has also been assumed that it takes about 10 years for the complete development of the disease to be diagnosed. It is clear, therefore, that at our present understanding of the pathogenesis, early diagnosis may allow preventive measures to be taken early enough to arrest, or halt the progression of the disease to a full-blown picture. Utsinger has reported that the likelihood of patients to exhibit the complete spinal manifestations of the condition increases with age. Some patients with solely peripheral enthesal involvement later developed the characteristic spinal picture, although the time elapsed from the first observation of peripheral enthesopathies to definite spinal DISH was not reported.

It is clear that peripheral enthesopathic involvement in DISH is common, and is often the promoter for ordering appropriate spinal radiographs that eventually lead to its diagnosis. The same concept can be applied to hypertrophic osteoarthritic changes, particularly if atypical sites are involved. Can DISH be limited to the peripheral joints and annexed soft tissues? Probably yes; however, at present we lack measures to establish such a diagnosis. Because most, if not all, research into the pathogenesis of DISH involves patients with established disease, no knowledge has been gained on metabolic, inflammatory, or enthesal changes in the early phases of the disease. It is therefore important to establish new diagnostic criteria that will take into consideration not only the radiographic aspects of the thoracic spine, but will encompass the clinical manifestations, the distribution and features of peripheral joints and enthesal sites involved, and aspects of spinal involvement other than the T-spine. Until that happens, we will diagnose the condition in its fully developed and probably irreversible phase, rather than its early and hopefully manageable phases.

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REFERENCES

Figure 1. An exuberant joint capsule enthesopathy of a 59-year-old man with no evidence of spinal DISH.

Mader: Editorial

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