

# Case Resolution

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*Presentation of the case on page 71.*

## Diagnosis

Diffuse idiopathic skeletal hyperostosis.

## Discussion

Diffuse idiopathic skeletal hyperostosis, DISH, is an idiopathic form of degenerative osteoarthritis which typically affects >60 year-old individuals (with an average prevalence of about 10% in >50 year-old people), and affects more frequently males than females. It is a disorder with no geographical preferences.

It is characterized by ossification of ligaments and entheses, especially in the axial skeleton, but it can also affect peripheral joints. In the vertebral column, it usually shows as exuberant bone growth along the anterior longitudinal ligament.

Previously acknowledged as the Forestier’s disease, senile ankylosis or ankylosing hyperostosis, DISH was described for the first time by Forestier and Rotes-Querol in 1950. These authors describe ossification (with “candle wax” appearance) along the anterior longitudinal ligament and the anterior-lateral surface of consecutive vertebrae (especially at the level of the thoracic vertebral column) with likely fusion of vertebral bodies. It had been described numberless times and it had been named different names, but it was only classified as an individual entity in 1950.

Although DISH, as it is suggested by its name, is an idiopathic condition of unknown etiology, there are reports on different causing agents such as diabetes mellitus, obesity, old age, hyperuricemia, HBP, hyperinsulinemia and insulin-like growth factors. Some studies also suggest traumatism, occupation, fluorosis, infectious diseases, autoimmune reactions (HLA-B27, HLA-B5, HLA-A11), neuropathies, and hypervitaminosis A as primary etiology. However, it has been decades since its relationships with social class, diet and life-style, obesity and diabetes has been proposed as the main triggering factor in this condition. There are paleopathology studies focused on highlighting the prevalence of DISH in ancient populations, relating it indirectly to life-style. These studies have evaluated the frequency of DISH among individuals in monastic and lay populations, and have suggested a pattern that shows higher frequency of DISH among individuals buried in monastic cemeteries, what suggests that those people who lived in monasteries (monks/priests) had a contributing factor distinctive of DISH. It has been suggested that this factor was a balanced and much richer diet and one with high contents of protein. These pieces of research try to illustrate the association of DISH signs with individuals who are properly fed or overfed.

At molecular level, this condition might be associated with genetic, metabolic, mechanic and vascular factors, and also with changes in signaling pathways such as Wnt, kB nuclear factor, BMP2, PG12 and endothelin 1. There have been studies on the prevalence of DISH in different world populations, and it oscillates between 2.9% and 42%, depending on racial and ethnic diversity and variations in the types of images and classification criteria that have been used. DISH prevalence is expected to increase in the next decades as a result of a higher life expectancy together with a global increase in metabolic syndrome.

Since in general it is painless and asymptomatic, DISH is more often than not diagnosed incidentally by a radiographic evaluation carried out due to unrelated causes. If there are clinical manifestations, symptoms are pain, restrictive lung disease, dysphagia, obstruction of upper airway, and skeletal rigidity. Vertebral column ankylosis in patients with DISH increases fourfold the risk of spinal fracture. Occasionally there can be pain at peripheral entheses sites.

In most patients, DISH can be diagnosed in simple X-rays and, if such is not the case, it is possible to get additional characterization of the condition in sectional images such as CT scan or MRI images. In general these ones are resorted to in trauma evaluation and the rare cases of spinal stenosis in patients with DISH caused by the ossification of the spinal ligaments or the hypertrophy of the interapophyseal joints.

In radiographic terms, there are findings in both the vertebral column and distant locations.

At the level of the vertebral column, disorders prevail between the 4<sup>th</sup> and 7<sup>th</sup> vertebrae. Usually ossification is located on the anterior or the antero-lateral aspects of the vertebral bodies and it is extended over the interbody spaces. There are degenerative changes in cervical apophyseal joints, ossification at nuchal ligament level, posterior osteophytes and calcification or ossification in the posterior longitudinal ligament. Moreover, there can be hyperostosis in the atlantoaxial joint and the occiput.

The thoracic vertebral column is the most frequently affected region in DISH, hyperostosis prevailing in the middle-lower region (7<sup>th</sup>-11<sup>th</sup> vertebrae). Ankylosis is most frequently seen in the thoracic column than it is in the cervical or the lumbar vertebral column. Although ossification can affect either the right or the left side of the vertebral bone, it is most frequent on the right side, what is attributed to the ossification inhibiting effects of the descending aorta pulse. In fact, patients with inversus situs develop ossification on their left side.

Moreover, there are radiolucent areas within bone ossification at interbody disc level. Ossification on the posterior aspect of the thoracic vertebral column is rare.

DISH at the level of the thoracic vertebral column can be accompanied by cortical thickening and hyperostosis on the posterior aspect of the ribs. However, apophyseal joints and costovertebral joints are preserved and do not get fused in this condition.

In the lumbar region, DISH more frequently shows as exuberant ossification from the zone proximal to the vertebral body anterior-superior angle ("spark plug flame" image). Ossification can be from 1 to 20 mm-thick.

The upper lumbar segments are the ones more frequently affected. Radiographic anomalies along the anterior aspect of the lumbar vertebral column are similar to those in the cervical column. Differently from the way it is in the thoracic vertebral column, ossification is equally frequent on the right and left sides of the lumbar vertebral column. There can be ossification upon the vertebral spinous processes and the interspinous ligaments. The degenerative narrowing of the interbody disc space is typically from mild to moderate. Degenerative changes in apophyseal joints can occur at the levels of the lower lumbar vertebral column and the lumbosacral joint, but there is no fusion.

At sacroiliac joint level, there can be bridge ossification over the anterior articular margins, what results in paraarticular fusion. Asymmetric intraarticular fusion, if any, prevails in the proximal fibrous portion of the sacroiliac joints.

The most used criteria for DISH diagnosis are the ones set out by Resnick and Niwayama:

1. The presence of exuberant ossification on the antero-lateral aspect of at least four consecutive vertebrae, forming bone bridges

2. The relative preservation of interbody spaces in the affected segment, and the absence of degenerative changes at disc level (vacuity or sclerosis of the vertebral body)

3. The preservation of the sacroiliac (erosions, sclerosis, reduction of intraarticular space) and the posterior inter-apophyseal joints.

The two first criteria help differentiate DISH from spondylosis, and the third one, from ankylosing spondylitis. Occasionally differential diagnosis is difficult because both conditions can coexist.

The classification criteria defined by Julkunen are almost equal to those set out by Resnick and Niwayama, except that they include the bone bridges that connect at least two vertebral bodies in two different sites of the thoracic vertebral column. Neither of these sets of criteria evaluates peripheral findings in this condition, though. The DISH criteria defined by Utsinger decreased the number of affected vertebral bodies to three consecutive vertebral bodies, but added the presence of peripheral enthesopathy. However, all the DISH classifications in specialized literature have limitations, especially because they describe advanced stages, and consensus in DISH classification criteria is still to be reached.

Distant disorders are very frequent and characteristic. Although they can show at any location, they are typically in the pelvic bones, the hip, the calcaneus and the knee. In the pelvic bones, hyperostosis prevails in almost 70% of the cases and consists of an increase of density at iliac bone level, irregular periostosis on the edges of the iliac and ischium bones, ossification of iliolumbar and sacroschial ligaments, and bone bridges on the lowest edges of the sacroiliac joints or the upper part of the pubic symphysis.

In the hip there is acetabular ossification, which can be differentiated from osteophytes in degenerative hip osteoarthritis by its wide implantation base and its gross aspect; square or triangular in shape and with irregular borders, it is usually located in the upper-external or lower-internal region of the hip cotyle ridge. The joint line is usually preserved. However, similarly to what has been described for the spine, it is not infrequent that these disorders coexist with hip osteoarthritis findings and, in such case, there may be joint line narrowing that makes diagnosis difficult.

In peripheral joints, hyperostosis is not as frequent as that. Incidentally hyperostosis may be diagnosed in the patella, where it originates on the anterior patellar aspect (at the level of the quadriceps insertion) as opposite to osteophytes in knee osteoarthritis, which lay on the femoral aspect. There may also be hyperostosis in the ankle (calcaneus posterior aspect) and the elbow (triceps olecranon insertion).

CT scan is not usually indicated in DISH, unless it is necessary to evaluate complications such as fracture, spinal canal stenosis secondary to the ossification of the posterior longitudinal ligament, or in case compressive effects on the esophagus or the inferior vena cava are suspected.

CT scan shows ossification along the anterior aspect, and reconstruction in coronal and sagittal planes usually shows the classic pattern of ossification and bone bridges.

The excellent spacial layout provided by CT scan allows evaluators to carry out precise assessment of facet joints, what is important to differentiate DISH from ankylosing spondylitis.

In general, MRI is not initially indicated in DISH either. However, it can be of value when the ossification of the posterior longitudinal ligament causes neurologic symptoms, because it allows evaluators to determine the extension of ossification, the mass effect on the thecal sac and the presence of compression on the spinal cord. Typically DISH shows as a long segment of low T1 and T2 signal which is in front of several consecutive vertebrae, whereas the ossification of the posterior longitudinal ligament shows in the same way behind the vertebral body and gets extended along several segments. The spinal cord edema shows as a diffuse increase in the T2 signal in the spinal cord parenchyma.

MRI might detect early changes in the axial and the peripheral skeleton. In this sense, recent MRI studies (and other US studies) suggest a local inflammatory process in affected entheses which might anticipate the process of ossification.

## Conclusions

DISH is a non-inflammatory condition which implies exuberant calcification and ossification in the anterior longitudinal ligament of the vertebral column and that of multiple peripheral entheses. It affects all the vertebral column, although it affects preferably the thoracic segment.

Differently from the impressive structural disorders that characterize other conditions, DISH can be largely asymptomatic. This is another reason because of which this condition has not been sufficiently paid attention to by medical research studies—because of the difficulties it presents for early diagnosis and appropriate treatment.

However, different clinical studies have confirmed the role that DISH plays as a disorder associated with many other systemic conditions, such as underlying metabolic disorders or cardiovascular disease.

Nowadays imaging studies are the most frequently used methods to consider DISH as a likely diagnosis, because DISH clinical symptoms are usually scarce and there are not specific lab tests either.

Characteristic findings in conventional radiology in DISH are the formation of new bone along the antero-lateral aspect of the vertebral column forming bridges between at least four consecutive vertebral bodies and in the absence of degenerative disc disease and inflammatory disorders in the sacroiliac and facet joints, what differentiates DISH from the degenerative spinal disease (spondylosis) and ankylosing spondylitis.

When there are symptoms, especially neurologic symptoms (such as those likely to occur in ossification of the posterior longitudinal ligament), it may be necessary to conduct more thorough studies with CT scan and MRI images.