Abstract:
Spinal involvement in alkaptonuria is common. Alkaptonuria with canal stenosis and instability has however not been reported. We wish to report one such patient. Lumbar canal stenosis with instability following ochronosis is a disorder with limited reports in the literature. This case is being presented for the surgical management of one of the complications of alkaptonuria- ochronotic spine. A 32 year old male was referred from the Rheumatology department with a diagnosis of Alkaptonuria. The patient had severe back pain and was unable to stand or sit or walk for some distance or lie down comfortably in bed. X-ray of the Lumbar spine revealed loss of lumbar lordosis, gross narrowing of the disc spaces, dense calcification of all the discs and advanced osteoporotic and degenerative changes. Patient also demonstrated instability in the lumbar spine. These findings confirmed that the patient was having an ochronotic spine. MRI-LS spine showed severe canal stenosis at multiple levels. Patient was taken up for decompression surgery to relieve his symptoms. In addition an instrumented posterolateral fusion was done to address the instability. The spine was so osteoporotic that pedicle screws had poor purchase in the vertebra. To overcome this problem, a spino-pelvic fixation was done on the right side. At follow up after 12 months, patient is comfortable being able to sit or stand comfortably and walk for long distances. However he still has pain in the lower back due to osteoporosis. He is on treatment for severe osteoporosis.

Keyword : Ochronotic spine, stenosis with instability, posterolateral fusion
INTRODUCTION:
Alkaptonuria is a rare autosomal recessive metabolic disease in which homogentisic acid, an intermediary product in the metabolism of phenylalanine and tyrosine cannot be further metabolised due to lack of the enzyme homogentisic acid oxidase. The accumulation of homogentisic acid leads to deposition of dark yellow, melanin like pigment in connective tissues leading to ochronosis. Homogentisic aciduria, ochronosis and arthritis are the characteristic triad of this metabolic defect. Patients with ochronosis present with spinal stiffness, peripheral arthritis, pigmentation of cartilage, sclera and skin.

CASE REPORT:
A 32 year old male was referred from the Rheumatology department with a diagnosis of Alkaptonuria. The patient had severe back pain and was unable to stand or sit or walk for some distance or lie down comfortably in bed for the past two years with preoperative Oswestry score of 41 - 60 % (severe disability).
Physical examination revealed a middle aged man of average built. Both pinnae of ears showed a few dark coloured spots. The pinnae were hard on palpation and deposition of ochre pigment in the conjunctiva was also noted. On questioning, the patient admitted that his urine would become dark coloured on prolonged standing. Examination of the lumbar spine revealed decrease in lumbar lordosis with restricted movements of the lower back. The cervical spine showed no significant abnormality. X-ray of the Lumbar spine revealed loss of lumbar lordosis, gross narrowing of the disc spaces, dense calcification of all the discs and advanced osteoporotic and degenerative changes. Patient also demonstrated instability in the lumbar spine. These findings confirmed that the patient was having an ochronotic spine.
MRI-LS spine showed severe canal stenosis at multiple levels.

Loss of lordosis
flexion-extension views demonstrating
L3-L4/ L4-L5/L5-S1 instability
And a diagnosis of **Ochronotic spine with L3- L4, L4-L5 & L5-S1 instability and L3-L4 & L4-L5 canal stenosis** was made. Patient was taken up for decompression surgery (L3 L4 L5 - laminectomy) to relieve his symptoms, as conservative treatment with bed rest and analgesics failed to alleviate his symptoms. In addition, an instrumented posterolateral fusion was done to address the instability. The spine was osteoporotic that pedicle screws had poor purchase in the vertebra. To overcome this problem, a spinopelvic fixation was done on the right side.

**RESULT:** pigment deposition noticed over interspinous ligaments
PECRULARITY OF THIS CASE:
Facet joint degeneration leading to root canal stenosis is a widely recognised pathological entity. However on reviewing the literature we have not found this described in Ochronotic Spondylosis. In addition, this patient had instability at L 3 – L 4, L4 – L 5 and L 5 – S 1 level. This patient had gross narrowing of the disc spaces, dense calcification of all the discs and advanced osteoporotic and degenerative changes. In contrast to commoner types of degenerative spinal disorders, vertebral osteophytes in ochronosis are poorly developed. We think the florid osteophytosis in this patient is the result of L 3 – L 4, L 5 – S 1 instability coupled with excessive stresses from the heavy manual work he was engaged in. MRI showed marked degenerative changes at the facet joints with prominent osteophytes encroaching into the spinal canal and IV foramina.

DISCUSSION:
Spinal involvement in Alkaptonuria is common. Ochronotic spine commonly presents with pain and stiffness. Lumbar canal stenosis with instability following Ochronosis is a disorder with limited reports in the literature. Osteoporosis is a feature associated in ochronotic spine. This case is being presented for the surgical management of one of the complications of Alkaptonuria-Ochronotic spine.

REFERENCES: