

# Kummell's disease: literature update and challenges ahead

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## Abstract

### Introduction

Osteonecrosis of the vertebral body, also known as Kummell's disease, occurs relatively rarely after vertebral compression fractures. However, as the life expectancy and resulting osteoporosis are on the rise, the incidence of the disease is bound to increase. We attempt to review the literature and discuss the future challenges about this not so well-known entity. The update is accompanied by a representative case report.

### Case Report

A 58-year-old physician presented with pain in mid-back region for a period of 15 days with a similar episode 5 months back lasting 3 weeks. Diagnosis of Kummell's disease was established on history pattern and radiographic examination. Anterior decompression and posterior pedicle screw fixation was performed in same sitting.

### Conclusion

A thorough knowledge of Kummell's disease and index of suspicion in appropriate situations is necessary for detection and discharge of effective treatment.

### Introduction

Kummell's disease (KD) refers to post-traumatic delayed collapse secondary to osteonecrosis of the vertebral bodies. Since its original description in pre-roentgenographic era by Hermann Kummell<sup>1</sup>, very few cases have been reported with

many not even completely fulfilling diagnostic criterion. Various terms have been assigned to denote the disease, such as vertebral pseudoarthrosis, intervertebral vacuum, cleft or gas, delayed vertebral collapse or vertebral compression fracture non-union<sup>2</sup>. The hallmark of the disease is the onset of back pain in a typically delayed manner, months to years after sustaining a trivial spinal injury.

Osteonecrosis of the vertebral body results due to the disruption of vascular supply by minor injuries. Osteoporosis is a contributory factor in spinal injuries, particularly low energy ones; consequently, low bone mass has become the most common cause of KD. Various studies<sup>3</sup> have shown the upward trend of osteoporosis, particularly in developing countries due to demographic transition and ageing population along with scarcity of resources. This can have profound implications on the incidence of fragility vertebral fractures. Fortunately, most of these injuries are relatively innocuous and heal without complications<sup>4</sup>; very few progress to develop osteonecrosis. However, some authors say that the incidence is quite high as opposed to that observed in clinical practices. The reason for this disparity has been suggested to be the varied terminology used<sup>2</sup>. We feel that the reasons are manifold poor reporting, no standardisation of definitions and, important yet unfortunate, lack of knowledge on part of attending clinicians. This report presents a literature review, which describes a prototypic case and discusses the challenges which lie ahead in the future.

### Method

We have searched the available literature on KD through Pubmed/Medline. Around 18 reports<sup>2,5-21</sup> were

identified which claimed to report a case of KD. Individual cases were analysed in detail (Table 1), and the reasons for exclusion/inclusion were also tabulated. Ten cases could fulfill the criteria necessary for KD. Collective analysis inferred from the data have been mentioned at appropriate places in the review.

### Historical description

In 1891, Hermann Kummel first gave the description of the disorder in six patients<sup>1</sup>. He described three stages of the disease: first, a stage of acute trauma followed by an asymptomatic period; secondly, a stage of recurrence of back pain and the last stage being appearance of kyphosis and neurological deficit. Radiographs had not been developed at that time, and the concept of initial normal radiographic picture was added many years later<sup>5,22</sup>. Steel<sup>5</sup> further elaborated the chronology of the events and suggested five stages beginning with the initial trauma, phase of minor pain, asymptomatic period, onset of severe pain and kyphosis and finally spinal cord compression.

Young et al.<sup>9</sup> searched English literature after 1950 and found that only five cases met Kummell's original criteria for the diagnosis among many reported. However, in many instances, the classical radiographic chain can not be completed because of the absence of post-traumatic initial negative radiographs. This is especially true in developing countries where either out of ignorance or poor resources, initial medical care is not sought at all.

### Aetiopathogenesis

Kummell<sup>1</sup> proposed that the nutrition status of the affected vertebral bodies is injured by the slight initial

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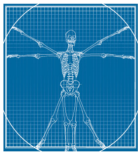


Table 1 Summary of cases of KD reported in the literature (including the case presented here)												
S.no.	Author	Year reported	No. of cases reported	Age (years)	Sex	Vertebral level	History of trauma	Asymptomatic period	Predisposing factor	Treatment	Surgical indication if surgery performed	Remark (if not fitting the description of Kummell's disease)
1	Steel	1951	1	23	Male	D10	Present	6 months	None	Conservative	-	-
2	Steel	1951	1	62	Male	D8	Present	6 months	None	Conservative	-	-
3	Brower	1981	1	71	Male	D12	Present	3 weeks	Osteopenia	Not mentioned	-	-
4	Hermann	1984	1	45	Female	L1	Present	1 month	Gaucher's disease	Not mentioned	-	-
5	Eanenaam	1993	1	75	Male	D11	Present	3 weeks	Steroid intake	Conservative	-	-
6	Young et al.	2002	1	72	Male	L4	Shovelled snow	6 weeks	None	Operative—corpectomy and global fusion	Neurologic deficit present	-
7	Chou et al.	1997	1	69	Female	L4	None	NA	Osteopenia	Operative—resection and fibular strut graft	Radiculitis present L4	No history of trauma or asymptomatic period
8	Freedman et al.	2009	1	78	Male	L3	None	NA	Osteoporosis	Posterior decompression and open kyphoplasty	Neurogenic claudication, sterile psoas abscess	Exacerbation of chronic symptoms only
9	Giraldo et al.	2012	1	45	Male	D12	Mild exertion	None	HIV infection, osteoporosis	Conservative, HAART modification	-	Continuous symptoms since initial episode; no asymptomatic period
10	Hur et al.	2011	1	73	Female	L1	Fall	8 years	Osteoporosis	Kyphoplasty	Claudication	-
11	Kim et al.	2011	1	82	Female	L1	Fall	None	Osteoporosis	Vertebroplasty	Severe pain, epidural haematoma	Continuous symptoms since initial episode; no asymptomatic period

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**Table 1 (Continued)**

12	Kim et al.	2012	1	72	Male	L1	Fall	14 months	Osteoporosis	Cement-augmented posterior instrumentation	Severe pain, posterior element involvement	Osteoporotic collapse present at the time of initial trauma too
13	Lee et al.	2008	1	72	Female	D12	None	NA	Mild osteoporosis	Decompression, vertebroplasty and posterior instrumentation	Epidural haematoma, paraplegia	No history of trauma or asymptomatic period
14	Ma et al.	2010	1	75	Female	D12	Fall	4 months	None	Vertebroplasty	Severe pain	-
15	Matzarglou et al.	2010	1	31	Male	L1	Repetitive flexion loading (construction worker)	1 year	None	Vertebroplasty	Severe pain	-
16	Fabbriciani et al.	2012	1	81	Female	L1	Fall	None	Osteoporosis	Osteoanabolic therapy	-	No asymptomatic period; initial radiographs showed normal spine
17	Osterhouse et al.	2002	1	79	Male	L2	Twisting	None	Osteopenia; chronic steroid intake	Referred; further treatment not mentioned	-	No asymptomatic period; initial radiographs showed normal spine
18	Schaaf et al.	2009	1	87	Female	L1	Minor trauma	Several months	None	Vertebroplasty	Severe pain	-
19	Swartz et al.	2008	1	60	Male	D9,D10	Fall	None	Diabetes mellitus with polyneuropathy	Corpectomy with posterior instrumentation and anterior graft filled cage	Pain, neurological deficit	Continuous symptoms since initial episode; no asymptomatic period
20	Present report	2013	1	58	Male	D12	Fall	5 months back	Osteoporosis, psychiatric illness	Corpectomy with global fusion	Pain, kyphosis	-

Competing interests: none declared. Conflict of interests: none declared.  
All authors contributed to conception and design, manuscript preparation, read and approved the final manuscript.  
All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.



trauma, so that softening and resorption of the vertebrae occur resulting in a late collapse. The collapse most commonly occurs at the junction of anterior one-third and posterior two-thirds. This is an area, which was proved by angiographic studies, represents a watershed zone of circulation<sup>10,23,24</sup>. Loading of the spine in hyperflexion has been described as the most common inciting initial cause; however, the majority had an underlying predisposing vertebral disorder, with the most common being osteoporosis. About 55% of the cases reported had low bone mass as a predisposing factor. Prolonged steroid intake also predisposed to the disorder in two cases by virtue of fatty infiltration, which occludes the intramedullary circulation<sup>8,19</sup>. Osteoporotic vertebral bodies are more prone to develop KD due to distortion of trabecular microarchitecture and the consequent fragile nature of the bone. The most common mechanism of occurrence of initial trauma has been observed in elderly, but one report<sup>17</sup> described occurrence of the disorder in a young adult male without any frank trauma.

### Clinical and Radiologic Features

The most common age group affected is elderly population, with a slight male preponderance<sup>5</sup>. The mean age at affliction was 65.5 years (range 23–87 years). Thoracolumbar junctional area is the most commonly affected, being a mechanical transition zone between rigid thoracic and mobile lumbar spine<sup>25</sup>. We found that 60% of the cases occurred between the eleventh dorsal and first lumbar vertebrae. Numerous microfractures occur ordinarily even in the absence of trauma in this zone. The classical clinical presentation of KD is vertebral body collapse, which occurs in a delayed manner after an initial trauma. The mean asymptomatic interval in cases where an initial trauma was present came out to be around 13 months with the longest being 8 years<sup>10</sup>.

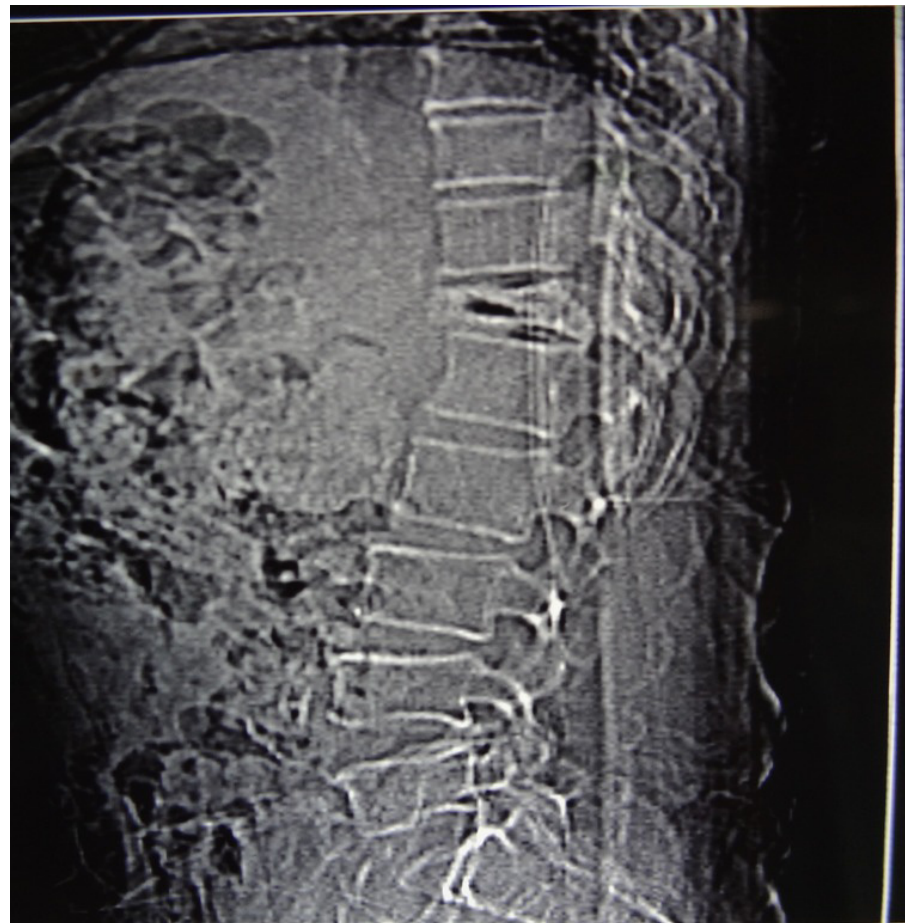
Serial radiographs consisting of initial normal ones and the recent ones showing vertebral collapse are pathognomic of KD<sup>6</sup>. Intravertebral cleft phenomenon was described by Maldague et al.<sup>23</sup>, according to whom the finding was pathognomic of KD. However, intravertebral gas appearance on plain radiographs and CT can also be encountered in neoplasia, infection, osteoporosis, chronic steroid intake, radiotherapy, intraosseous disc prolapsed, arteriosclerosis, alcohol abuse, pancreatitis or cirrhosis<sup>26</sup>. Magnetic resonance imaging (MRI) scan confirms the presence of air fluid levels in the vertebrae and may present a classical double line sign<sup>9</sup>.

As this is a rare disease, all other causes of vertebral collapse should be ruled out before considering KD.

MRI scan should be obtained in all cases to rule out perivertebral inflammatory changes. Haematological and other relevant investigations should be carried out to adequately rule out malignancy. Some cases might still require a biopsy if the diagnostic work up is ambiguous.

### Treatment

Non-surgical treatment for KD consists of braces and analgesics. However, the efficacy of such a treatment is debatable<sup>27</sup>. As the natural history of the disease ends in vertebral body collapse, kyphosis and spinal cord compression, it is prudent to stem the course and prevent such consequences. We feel that it is important to understand here that the treatment of this entity is different



**Figure 1:** Plain lateral radiograph showing collapse of D12 vertebrae with intravertebral gaseous shadow.

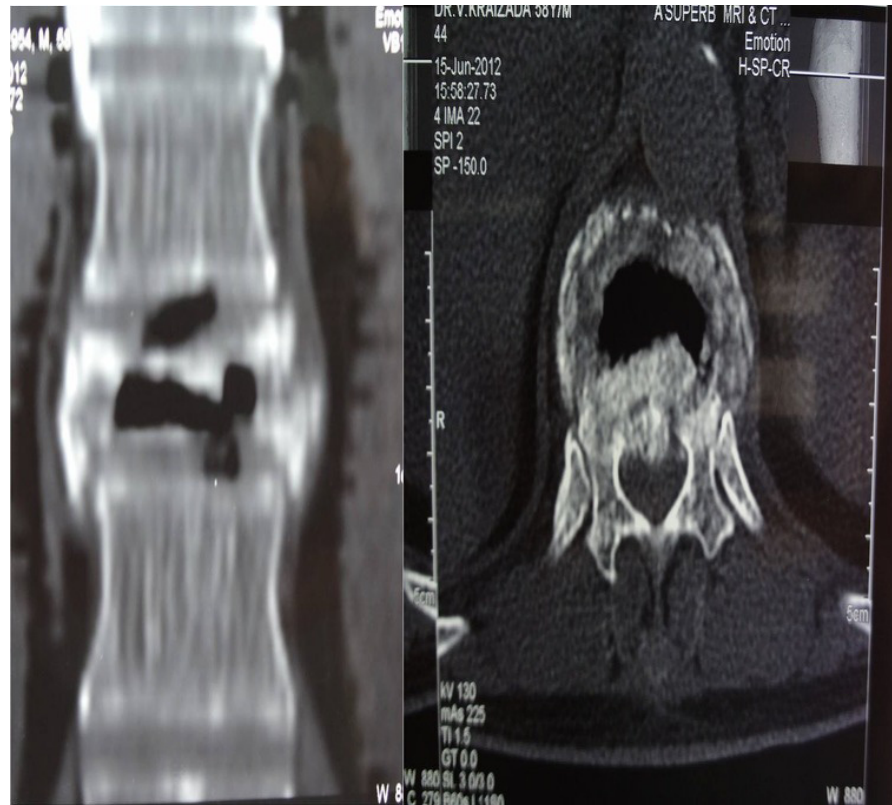
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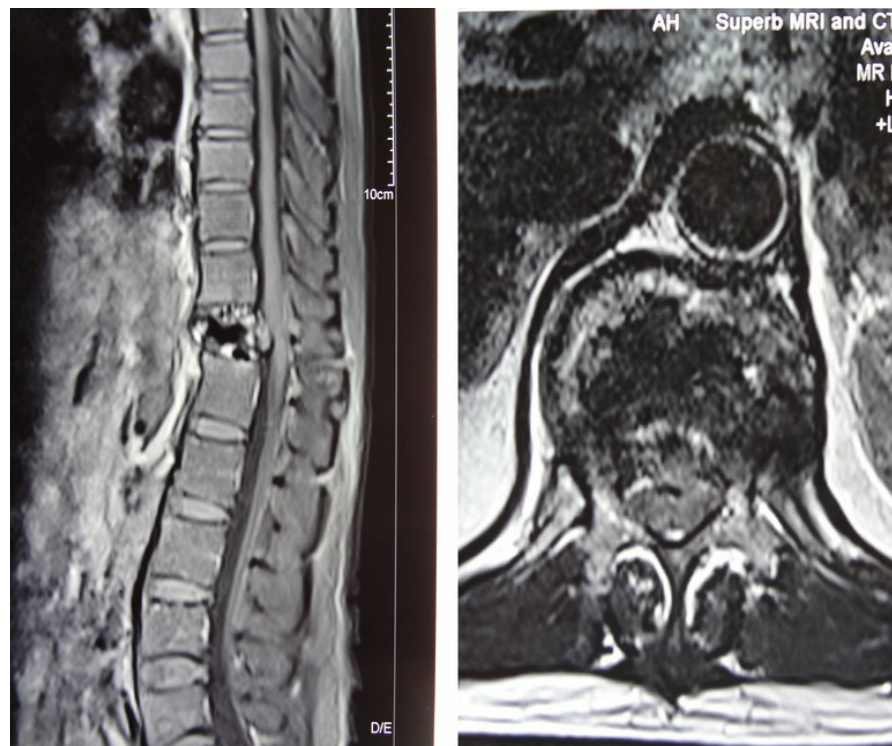
from osteoporotic vertebral collapse because the damage in the former is ongoing in the form of osteonecrosis. Recently, Fabbriciani et al.<sup>18</sup> evaluated the role of osteoanabolic therapy in the form of teriparatide in an 81-year-old female and found the outcome to be satisfactory. In our opinion, such form of treatment should be reserved only for patients with medical comorbidities, i.e. who are high-risk candidates for surgical intervention. Although different in indications, same principles of management, as applicable to osteoporotic collapse, apply to the treatment of KD as well. Surgical intervention for KD can be decided on the basis of Li's staging of the disease—stage I: vertebral body compression <20%; stage II: vertebral body compression >20% with adjacent disc involvement; and stage III: with posterior cortex involvement with spinal cord compression. They advocated augmentation alone in stages I and II, and decompression and surgical stabilisation in stage III<sup>28</sup>. Augmentation by vertebroplasty or kyphoplasty is an effective means of alleviating pain<sup>29</sup>; however, a spinal cord compression requires a formal open procedure<sup>14</sup>. The choice between an anterior and posterior procedure is based mainly on the surgeon's discretion; but in most of the cases, a global fusion is required due to the presence of concomitant osteoporosis.

### Case Report

A 58-year-old physician came with complaints of pain in the back and kyphotic deformation of the thoracolumbar junction for the last 15 days. There was no neurological involvement, no history of definite trauma, fever, night chills or constitutional symptoms. He had a past history of psychotic illness, for which he was taking haloperidol. His son recalled he had a fall following a hyponatremic episode 5 months back. The pain persisted for about 3 weeks at that time.



**Figure 2:** CT scan images of sagittal, coronal and axial cuts showing intravertebral cleft and mild retropulsion.



**Figure 3:** MRI images showing lack of perivertebral inflammation, high signal intensity on T2W axial cut.

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**FOR CITATION PURPOSES:** Jindal N, Sharma R, Jindal R, Garg SK. Kummell's disease: literature update and challenges ahead. *Hard Tissue* 2013 Dec 30;2(5):45.

Competing interests: none declared. Conflict of interests: none declared. All authors contributed to conception and design, manuscript preparation, read and approved the final manuscript. All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.



Radiographs were obtained, which showed collapse of the 12th thoracic vertebra with intravertebral gas shadows (Figure 1). CT images showed a heterogenous pattern of gas within the vertebral body (Figure 2). Mild retropulsion of the posterior vertebral body was observed in axial cuts. MRI scan (Figure 3) confirmed the presence of air in the vertebra with no adjacent inflammatory changes. A high-intensity band was visible on T2-weighted images confirming the presence of avascular necrosis. Tests for infection, metastasis and multiple myeloma were negative. Bone mineral density revealed severe osteoporosis with a *t*-score of -3.6. Flexion extension radiographs did not show significant movement at the non-union site.

Owing to cord compression and severe kyphosis, a surgical plan of decompression and anterior strut graft was made. Posterior pedicle screw

instrumentation was carried out first followed by anterior corpectomy and cage fixation, in the same sitting (Figure 4). The post-operative period was complicated by acute delirium, which settled after 3–4 days. Even after 15 months of the procedure, the patient has been mobilising comfortably without further complaints.

### Discussion

Freedman et al.<sup>2</sup> proposed that the incidence of the disease is quite high at 7–37%, especially in elderly age group. Fabbriani et al.<sup>18</sup> also advocated the probability of the disease in patients with chronic spinal symptoms, especially with osteoporosis. This coupled with the increasing trend of osteoporosis, especially in developing countries<sup>3</sup> is bound to increase the incidence of KD. The information regarding this entity is very little with many major orthopaedic texts<sup>30</sup> omitting the issue altogether.

We interviewed 15 orthopaedic residents in our institute, and found that only two of them knew of the disease and that too only superficially. Even spinal surgeons are not well-versed with this disorder, particularly with the management.

### Conclusion

Elderly individuals with spinal pain, especially in the thoracolumbar area with negative radiographs, should be followed up diligently to watch out for late collapse. The reverse is also true; patients with collapse should be carefully assessed for any traumatic event earlier and an asymptomatic period in between. In our view, this is the only differentiating feature from an old osteoporotic collapse, which has a far less malignant course and requires a different management strategy. However, the possibility of development of osteonecrosis in an already collapsed vertebra cannot be ruled out, although such a scenario would not fulfill the Kummell's criteria. The development of disproportionate pain in an osteoporotic vertebral collapse should alert the treating doctor to such an entity. A thorough knowledge of KD and index of suspicion in appropriate situations is necessary for detection and discharge of effective treatment.

### Abbreviations list

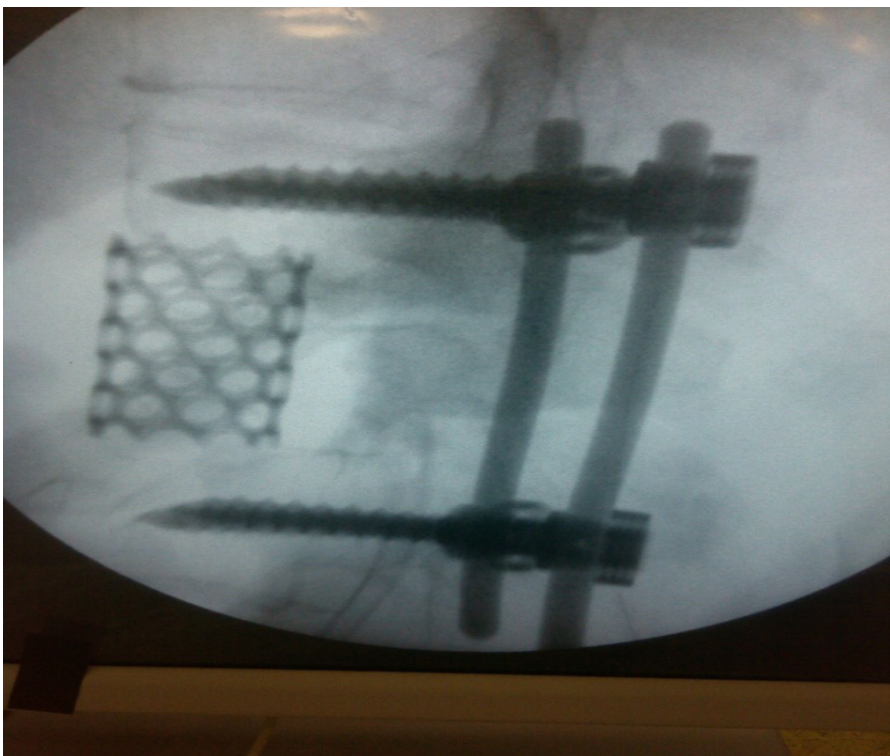
KD, Kummell's disease; MRI Magnetic resonance imaging.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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**Figure 4:** Final fluoroscopic image showing restoration of normal spinal curvatures.



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