A Systematic Review of Developmental Lumbar Spinal Stenosis

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DECLARATIONS

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ABSTRACT

Purpose

To systematically evaluate any consensus for the etiology, definition, presentation and outcomes of developmental lumbar spinal stenosis (DLSS).

Methods

A comprehensive literature search was undertaken by 2 independent reviewers among PubMed, Ovid, and Web of Science to identify all published knowledge on DLSS. Search terms included "developmental spinal stenosis" or "congenital spinal stenosis" and "lumbar". The inclusion criteria were English clinical studies with sample size larger than 8, articles examining the etiology, diagnostic criteria, surgical outcomes of DLSS, and its association with other spinal pathologies. Articles that did not specify a developmental component were excluded. The GRADE approach was used to assess their quality of evidence.

Results

The initial database review found 404 articles. 20 articles with moderate to very low quality met the inclusion criteria for analysis. The canal size was significantly smaller in patients with DLSS than normal subjects. In addition, the risk of re-operation on adjacent levels (21.7%)

was high which could be explained by multi-level stenosis. However, there was a lack of consensus on the methodology of diagnosing DLSS and on its specific surgical techniques.

Conclusion

Multi-level stenosis and re-operation are especially common with DLSS. Identification of these individuals provides better prognostication after surgery. However, current literature provides few consensus on its definition and the required surgical approach. Besides, there are limited reports of its etiology and association with other spinal pathologies. Due to these limitations, standardizing the definition of DLSS and investigating its etiology and expected clinical course are necessary.

Keywords

Developmental spinal stenosis; lumbar; magnetic resonance imaging; axial; bony spinal canal diameter

INTRODUCTION

Developmental lumbar spinal stenosis (DLSS), also known as congenital lumbar spinal stenosis, describes a pre-existing narrowing of the bony spinal canal. On the contrary, degenerative lumbar spinal stenosis refers to the cause for neural compression including disc herniation, spondylolisthesis, ligamentum flavum hypertrophy and facet joint osteophytes. It is important to differentiate them as they have different etiologies and their management is different. However, these two subtypes are not always mutually exclusive, as many degenerative LSS surgical cases have concurrent developmental stenosis[1,2]. Both pathologies indicate a pathoanatomical phenotype for canal size or compressive elements but clinically they are indistinguishable due to the common presentation of nerve compression.

DLSS was first illustrated by Verbiest in 1954 as narrowing of the spinal canal in the lumbar region with concurrent neurogenic claudication, radicular pain, and motor weakness in the lower limbs[3]. He described a pre-existing narrowed spinal canal with a low threshold for neural compression. It was noted patients with a smaller midsagittal canal diameter tends to have a higher chance of chronic lower back pain[4]. In a normal-sized spinal canal, mild degeneration may not be sufficient to cause significant clinical symptoms.

Defining DLSS is important when managing a patient with neural compression. A patient with DLSS is prone to disease at multiple levels[5-7] and these apparent less severely compressed levels may also require decompression surgery. Lower threshold may be prudent due to the risk of re-operation[8-10]. However, current diagnostic definitions and clinical implications for DLSS are ambiguous. Therefore, this systematic review aims to determine any consensus regarding the etiology, definition and clinical course of DLSS, and its associations with other spinal canal pathologies.

MATERIALS AND METHODS

Literature Search Strategy and Selection Criteria

Literature search was conducted following the PRISMA statement[11]. A comprehensive search was performed using PubMed, Ovid, and Web of Science to identify articles related to the current knowledge of DLSS. Search terms included "developmental spinal stenosis" or "congenital spinal stenosis" and "lumbar". The inclusion criteria and exclusion criteria were described in Table 1 and Table 2. We included case-series and observational studies in this review as we expected a scarcity of literature related to this topic. A sample size of 8 or larger was required for inclusion since a cut-off of 9 or above excluded at least two articles from the review. Given the limited number of studies, we did not want to raise the sample size minimum any further. This was done understanding the risks of introducing selection bias and insignificant effect sizes with small sample sizes[12]. Two investigators remained independent in the search process before convening for final inclusion. Discrepancies were settled through discussion during full-text screening. The references of each included article were screened through for any other pertinent articles.

Data Extraction and Critical Appraisal

The main outcomes extracted included (1) etiology, (2) imaging phenotypes (Table 3), (3) relationship of DLSS with other spinal canal pathologies, and (4) surgical treatment and outcomes and surgical complications (Table 4). Details regarding each study's sample size, mean age of subjects, imaging modalities adopted, radiological definitions of DLSS, years of follow-up, surgical indications, operative procedures, methods of assessing outcomes, surgical outcomes and their complications were recorded if applicable. Quality of studies included was assessed by using The Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach[13,14]. Randomised trials were given high quality of evidence, while observational studies and case-series were given low and very low quality of evidence respectively. The quality of evidence was downgraded by one level according to the following criteria: inconsistency of results, imprecision of data, high probability of reporting bias, and limitation to study design. The quality of evidence was upgraded by one level for the following cases: strong evidence of association between independent variables and outcomes and evidence of dose-response gradient.

RESULTS

The search results were compiled in a PRISMA flowchart (Figure 1). The initial search yielded 404 articles with 195 from PubMed, 87 from Ovid, and 122 from Web of Science. After excluding 84 duplicated articles, a total of 320 studies were available for title and abstract screening. After applying the inclusion and exclusion criteria, 65 articles were eligible for full-text screening. A total of 20 studies met the final criteria and were included. They were published between May 1977 and November 2019. Quality of evidence assessment is shown in Table 3 and Table 4.

Etiology

Only 1 study was included for the etiology of DLSS. Cheung *et al*[15] conducted a genome-wide association study on 469 asymptomatic subjects and obtained axial magnetic resonance imaging (MRI) with serum DNA. DLSS were identified by axial MRI according to values published by the same author[1]. They found the most significant single nucleotide polymorphism (SNP) was 4kb from the ZNF704 gene ($p=4.33\times10^{-7}$) on chromosome 8 for L4. For L5, the most significant SNP was the DCC gene ($p=4.67\times10^{-7}$) on chromosome 18. Another significant SNP was rs3781579 ($p=8.21\times10^{-4}$) of the low-density lipoprotein receptor-related protein 5 (LRP5) on chromosome 11 which was essential in Wnt signalling pathway for bone development. It met the Bonferroni threshold for significance. They also proposed

L1-L4 were clustered differently compared to L5-S1, suggesting a different genetic predisposition pattern of multilevel involvement in DLSS.

Diagnostic criteria

Eleven imaging studies on the definition of DLSS were found (Table 3). Of these, five examined the role of MRI[1,2,16-18], one analysed MRI and plain radiographs[5], two explored plain radiographs[19,20], and three illustrated the role of computerized tomography (CT)[6,7,21]. Different phenotypes were explored, including using axial and midsagittal anteroposterior (AP) canal diameter at the vertebral body and disc level, canal and vertebral body cross-sectional area, and pedicle length. Sample size, sex, mean age and radiological findings of each study were listed in Table 3.

Cheung *et al*[1] found the axial AP canal diameter at the vertebral body level was the most predictive imaging parameter for DLSS based on the area under the receiver operating characteristic (ROC) curve (AUC) analysis for all lumbar levels on axial MRI (AUC: 0.66-0.84, p<0.030 to <0.001). They defined relative DLSS based on including 50% controls with the best sensitivity (30%-65%) and specificity (68%-93%). Critical values, which included surgical cases and none of the controls, were also defined with high sensitivity (97%-100%) and specificity (80%-90%).

Chatha *et al*[16] performed midsagittal T2-weighted MRI measurement of the AP vertebral canal diameter at each vertebral body and disc level. By assuming the subjects would follow a normal distribution, they considered measurements larger or smaller than 2 standard deviations from the mean to be outliers. The lowest cut-off limit of the AP vertebral canal diameter had a range of 3.8mm at L3-L4 disc space to 9.3mm at L1. After rounding off, the authors proposed the cut-off limit of the AP vertebral canal diameter for DLSS.

Kitab *et al*[2,17] conducted an MRI-based multivariate analysis on 709 patients with lumbar spinal stenosis. The authors divided the patients into two cohorts: those who had symptoms before 60 and those who had symptoms after 60. They suggested their findings challenged the belief that stenotic changes at L4-S1 were mainly associated with degeneration (Table 3). The authors concluded that subjects with a narrowed spinal canal could not only be attributed to degeneration. Developmental narrowed spinal canal was the basic characteristic that predisposed patients to clinical symptoms of LSS. Lumbar spinal stenosis should be defined as a developmental syndrome with superimposed degenerative changes.

Similarly, Singh *et al[18]* also compared the radiological structural differences between patients with DLSS and age- and sex-matched controls. Subjects with definite DLSS were diagnosed clinically by a senior author. However, the authors did not provide the rationale for identifying those with definite DLSS. Axial and midsagittal MRI were obtained, and several parameters were measured. The results were presented in Table 3.

Kitab *et al*[5] analysed MRI and plain radiographs to find for possible anatomic variations. They diagnose DLSS as patients younger than 50 with neurogenic symptoms for at least 2 months, and with minimum radiological degenerative manifestation. However, this degenerative manifestation was not clearly defined. Subjects with deformity or instability were excluded. They conducted multiple measurements on MRI and found there was a reduction in several imaging parameters in patients with DLSS (Table 3).

Cheung *et al*[19] analysed AP and lateral standing plain X-Ray to search for radiological definitions of DLSS. DLSS was defined by using previously published cut-offs[1]. Several radiological measurements were obtained on AP and lateral radiographs (Table 3). The authors reported the SBW:PW ratio had the largest AUC and proposed level-specific cut-off ratios (Table 3).

Mrówka *et al*[20] compared routing or tomographic X-ray with contrast examination to evaluate its diagnostic accuracy. A midsagittal canal diameter of less than 15mm was defined to be pathological by the authors. They found X-rays were inferior to contrast examination (Table 3). The authors concluded clinical signs of narrowing were not characteristics of DLSS.

Similarly, Postacchini *et al*[6] assessed the diagnostic accuracy of CT. A cut-off value of 13mm was used to diagnose DLSS. Multi-level stenosis was noted in half of the samples. The authors concluded CT was less accurate than water-soluble myelography for DLSS.

The same authors also analysed CT conducted with the same protocol as above[6] to search for anatomical variations between groups[7]. Several radiological measurements were obtained, and the results were presented in Table 3.

They found most cases had the shortest canal diameter at L4-L5. The authors also noted multi-level stenosis and reduced laminae length in all cases, whereas interpedicular diameter and the size of bony canal to the size of vertebral body ratio varied.

Akar *et al[21]* also used CT to compare the morphometric data between DLSS and degenerative spinal stenosis patients. DLSS was defined as AP canal diameter of 15mm or below at the vertebral body level under axial CT images. The authors obtained several measurements, and the results were presented in Table 3.

DLSS and other spinal canal pathologies

Two articles[22,23] studied the relationship between DLSS and other spinal canal pathologies. In a group of 34 patients who underwent surgical decompression for lumbar spinal stenosis, Cheung *et al*[22] excised ligamentum flavum (LF) during surgeries for histological examination. Subjects with DSS had negative correlations with LF thickness and the degree of LF fibrosis. A similar inverse relationship was also observed for the area of LF fibrosis in critical DLSS patients, while a positive correlation was observed for non-DLSS patients.

Soldatos *et al[23]* performed a retrospective study investigating the association between DLSS and degenerative changes of the lumbar spine in patients with DLSS and controls. They defined DLSS as a spinal canal of smaller than 14mm on at least one lumbar level under midsagittal MRI. Imaging parameters including annular bulge, annular tear, disc herniation, epidural lipomatosis, Schmorl's nodes, spondylolisthesis and pars defect were assessed. The authors found patients with DLSS had a higher incidence of annular bulges, disc herniations, annular tears and spondylolisthesis (P=0.001-0.012).

Operative Management, Outcomes and Complications

Six studies reported the outcomes of surgical treatment in DLSS patients (Table 4), in which 2 studies compared between patients with DLSS and degenerative LSS[9,24], and 4 studies only focused on DLSS cohorts[8,10,25,26]. Different surgical techniques and outcome assessments were used, but all authors focused on the difference between preoperative and postoperative symptoms as clinical outcomes. Overall, the success rate of complete clearance of symptoms by DLSS surgery was 65%-68%[9,10,25]. Sample size, sex, mean age, average time of follow-up, methods of diagnosing DLSS, choices of surgical interventions, assessment of outcomes, and surgical complications were listed in Table 4.

Reale *et al*[9] compared the surgical results in patients who underwent low lumbar myelography with water-soluble contrast medium. Fewer DLSS patients (62.2%) reported excellent or good outcomes than degenerative patients (73.7%). The authors also found preoperative symptoms of urinary disturbance (86% vs 100%) and Lasègue's sign (65% vs 92%) responded best percentage-wise in both groups. However, patients with DLSS had an overall poorer surgical outcome than degenerative patients.

Louie *et al[24]* conducted a retrospective study comparing the surgical outcomes between DLSS and degenerative spinal stenosis patients. DLSS was defined as a shorter pedicle and smaller cross-sectional area of the spinal canal than normal under lateral plain radiograph. By using Charleston Comorbidity Index Score (2.8±1.6 vs. 0.5±0.6; $p<0.001$) and American Society of Anaesthesiology Score ≥3 (52.8% vs. 11.1%; $p<0.001$), they concluded patients with degeneration had more comorbidities than DLSS. Postoperative results showed no statistically significant differences in the visual analogue scale and the Oswestry Disability Index between groups (P=0.117-641). The levels of symptomatic relief were also similar.

Lee *et al*[26] divided the patients into three pathological categories: (1) Concentric stenosis, (2) Sagittal flattening, (3) Abnormal articular processes. Multi-level stenosis was noted in 15 out of 16 patients. 5 patients had satisfactory results (50-75 points) from surgery, while 5 were unsatisfied (30-50 points). They found the unsatisfied cases were due to inadequate decompression, in which some patients were not recognized as stenotic cases. Overall, surgical treatment had a better result than non-surgical treatment (49.3 vs 34 points).

Similarly, Dai *et al*[25] classified DLSS patients into the same pathological categories as Lee *et al*[26]. The authors reported preoperative symptoms disappeared in 28 patients, who rated excellent; 13 had some backache remained, and they rated good or fair; 1 had reappearance of intermittent claudication 6 years after surgery. They concluded there was no significant difference between clinical results and canal diameters.

Verbiest[10] also analysed the outcomes of surgical intervention but reported better outcomes when compared to Lee *et al*[26]. The author reported 62 patients (68.1%) were completely relieved from preoperative symptoms, with sciatica as the most frequently resolved symptoms (94.4%), and 29 had persistent symptoms after decompression. The rate of recovery is the highest with pure absolute stenosis and lowest with pure relative stenosis. However, the author found no difference in canal sizes when comparing between groups.

One of the most common complications of operation on DLSS patients is re-operation[8-10]. Cheung *et al*[8] identified 235 patients who underwent decompression had levels of DLSS adjacent to the index operated level. 51 (21.7%) of these patients had to undergo re-operation at these levels. L4-L5 was the commonest level (77.4%) to have single-level decompression at index operation, and it was also the commonest level that required adjacent level re-operation. Besides, the risk of reoperation was lower after multi-level decompression in subjects with DLSS. DLSS at the adjacent segment, the number of operated levels, and the patient's age at index surgery were used in multivariate regression model to predict the likelihood of re-operation at an adjacent segment, and it correctly predicted 89.4% of the cases with an adjusted odds ratio of 3.93. The authors concluded DLSS is a poor prognostic factor and susceptible levels should be identified prior to the index operation with consideration of prophylactic decompression.

DISCUSSION

In patients with pre-existing narrowed spinal canals, mild degeneration is sufficient to cause compressive symptoms, leading to a significant impact on functioning and quality of life. It is important to identify cases of DLSS and provide suitable treatment, to reduce re-operation rates and maximize prognosis. However, to date, there is no clear definition and clinical implication of DLSS. Therefore, the objective of this review is to identify if there is any consensus regarding the etiology and definition of DLSS, associations with other spinal canal structures and its clinical course.

Based on only one paper that has described the etiology of DLSS, there is a paucity of literature in this area. The genetic etiology illustrated by Cheung *et al*[15] provided an early approach to identify people that may be at risk, which would allow close monitoring and follow-up. However, the results were not generalisable to other populations besides Southern Chinese. Future studies should extend to other ethnicities for broader application. Also, similar to degenerative LSS[27], it is worthwhile to propose a pathophysiology hypothesis for DLSS to have a clearer perception of the disease.

Several studies examined the radiological diagnostic criteria of DLSS without much consensus. In contrast, the parameter used was quite consistent. The canal size is consistently smaller in patients with DLSS[1,5,7,16,18,21,28], and they are prone to have vertebral canal narrowing in multiple spinal levels[5-8,26]. For research purpose, the majority of the studies utilized AP vertebral canal diameter instead of the cross-sectional area of the canal because it is more readily available and more convenient to obtain. It is important to note that DLSS and degenerative changes of the spine often coexist on the same patient[2,17,22,23]. Differentiation of the two pathologies is essential to appreciate the radiological phenotypes and predict possible pain generators in a patient but may be challenging without clear radiological definitions. Different cut-off values were proposed to define DLSS which added difficulty in studying the pathology. Although Cheung *et al*[1] used axial MRI to visualize the spine, the method of including 50% of the control to define relative DLSS generated low sensitivity but relatively high specificity, indicating the diagnostic test would have a large number of false negatives, which is not ideal. Chatha *et al*[16] made measurements in the midsagittal view, which are affected by the posterior curvature of the vertebrae[29] and diseases at the disc and endplate[1]. They are subjected to great variability which adds to the difficulty of defining DLSS radiologically. Furthermore, the positioning of patients during imaging varied from supine to lateral standing or is not reported in some studies, hence their accuracy cannot be compared. Based on the current evidence, we suggest the diagnostic criteria by Cheung et al[1] is the most suitable for patients with DLSS as they are subjected to less variability. Nevertheless, it is necessary to standardize with a large cohort as most of the studies presented here are flawed with small sample sizes.

Based on only two articles that analyse the association between DLSS and other spinal canal pathologies, we should expand our knowledge in this area. Cheung *et al*[22] assessed the association of LF thickness with area of fibrosis and canal diameter, but its pathophysiology has yet to be discovered. The relationship between DLSS and degenerative spinal changes were also investigated[23] but with small sample size. This is one of the aspects that worth studying in depth, and a larger population should be utilized to provide stronger evidence. The role of epigenetics may also be a direction for future research.

For operative management, the choices of surgical intervention varied from simple discectomy[25] to laminae and articular processes removal[8,9]·[24,26]. However, some authors[9,25] did not provide the rationale of choosing their choice of surgical techniques, which may limit their generalizability. Most literatures were of low or very low quality of evidence as they failed to compare surgery with placebo, no treatments, or sham surgery. Verbiest[10] noted the choice of surgical intervention was dependent on patients' radiological signs and presentation during operation. However, many authors treated DLSS as general LSS and omitted the presence of multi-level stenosis. Therefore, with the above variations, it is difficult to draw any conclusions to the recommended surgical technique. This is an area that should be investigated as the current mix of techniques generated great variability and yielded unpredictable surgical outcomes.

The assessment of surgical outcomes was prone to bias as most studies only addressed the change in pain response without objective assessments. The surgical outcomes were generally consistent even though different surgical techniques were used. This is reiterated by other systematic reviews[30,31] which suggested no superiority between decompression techniques for treating LSS. However, a major flaw of these studies was the lack of differentiation between DLSS and degenerative types, in which the outcomes may be variable. Only 65%-68% of the patients who received surgical interventions achieved complete remission of preoperative symptoms. When compared to a re-operation rate of 13.0% as reported for LSS[32], Cheung *et al*[8] proposed a larger rate of 21.7% in patients with DLSS. As multiple stenotic levels are common, pre-existing narrowed canals that are asymptomatic are tend not to be investigated or operated. Hence, these levels are more susceptible to neurological compromise even if mild degeneration of the spine is present. Reoccurrence of compressive symptoms after initial decompression is an indication for reoperation. The characteristic of multi-level stenosis of DLSS is a poor prognostic factor and any at-risk spinal levels may need to be addressed at the index operation.

One of the major concerns with this systematic review is the inclusion of case series. They are prone to selection bias when the authors only select the relevant cases to report, which may not represent the general population. In addition, the internal validity of case series is relatively low as there were no control arm for us to compare. However, given the limited amount of literature available, they are included into this review in order to provide a relatively more comprehensive view on the current knowledge of DLSS. Other limitations also arise during the selection of literatures. Language bias is introduced as we only included English literature. Selection bias is also introduced when we only screened through 3 major databases for selection of eligible studies.

CONCLUSION

The current available evidence suggests patients with DLSS have a smaller vertebral canal size with the involvement of multiple levels. The high re-operation rate can be explained by presence of multi-level stenosis. It is crucial for surgeons to identify the presence of DLSS radiologically prior to index surgery for better surgical planning. However, the current definition of DLSS is vague and there is a lack of agreement. Future research should aim to develop a standardized definition. More work is required regarding its etiology and association with other spinal canal pathologies to better understand the pathomechanism of the disease.

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FIGURE CAPTIONS

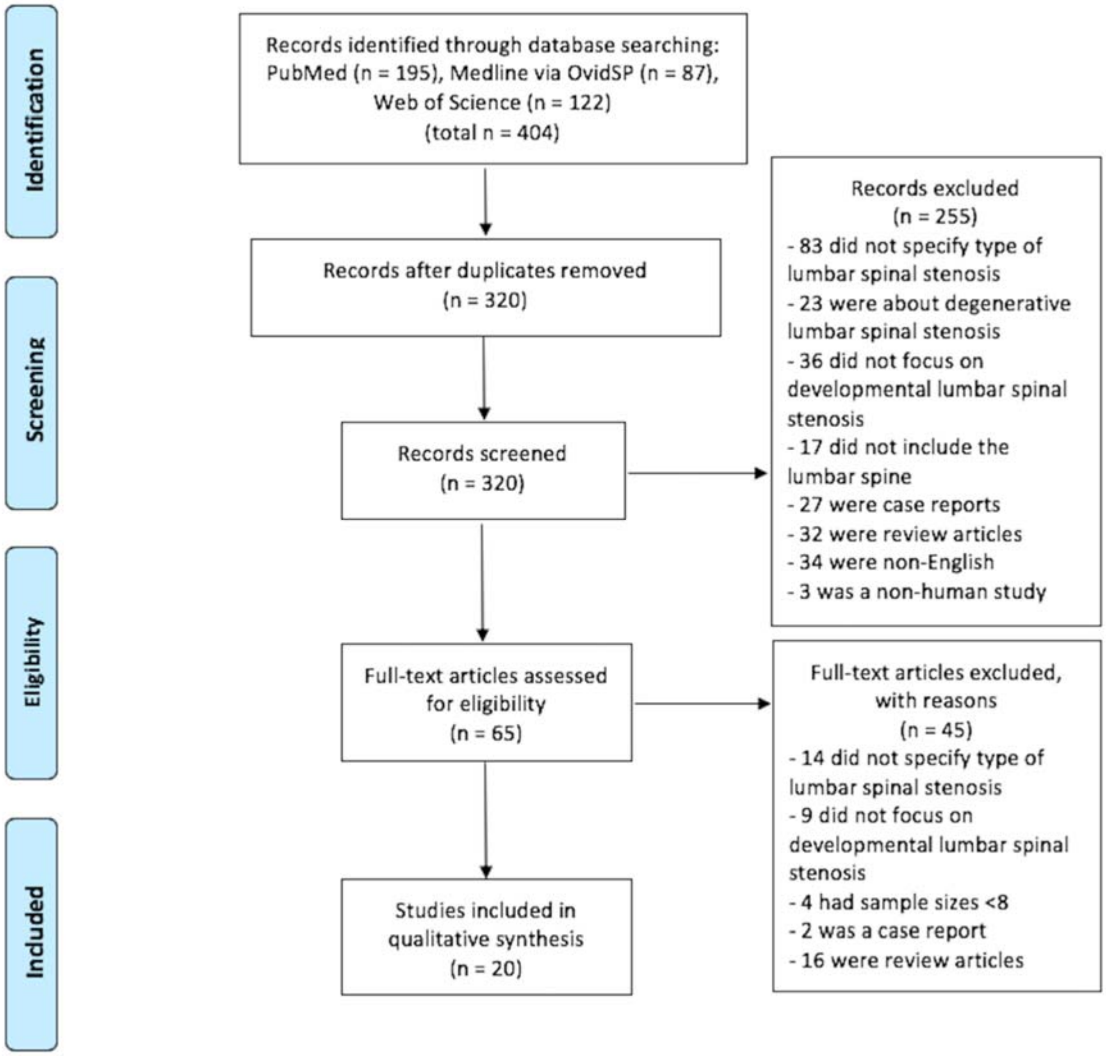

Fig1 Flowchart for Studies Included and Excluded in the Review

Table 1. Inclusion and Exclusion Criteria for Articles Investigating Imaging Definitions of Developmental Lumbar Spinal Stenosis (DLSS)

Inclusion Criteria	Exclusion Criteria
DLSS identified by magnetic resonance imaging, computerized tomography, or X-Ray	Non-English literature
Observational studies (cohort or cross-sectional or case-control study)	Case reports
Case-series with sample size of 8 or more	Animal studies
Randomised controlled trials	Systematic reviews and Meta-analyses

Table 2. Inclusion and Exclusion Criteria for Articles Examining Developmental Lumbar Spinal Stenosis treatments

Inclusion Criteria	Exclusion Criteria
Comparing preoperative and postoperative symptoms	Non-English literature
Methods of diagnosing DLSS were described	Case reports
Surgical techniques were described	Animal studies
Observational studies (cohort or cross-sectional or case-control study)	Systematic reviews and Meta-analyses
Case-series with sample size of 8 or more	
Randomised controlled trials	

Table 3. Studies that Examined Imaging Phenotypes of Developmental Lumbar Spinal Stenosis (DLSS)					
Study	Design	Quality of Evidence	Sample Size (Sex); Mean Age (Range) *(yr)*	Imaging Modalities	Radiological Findings
Cheung *et al*[1], 2014	Case-control study	Moderate	100 LSS surgical cases (48 M, 52 F) vs 100 age- and sex-matched controls (50 M, 50 F); 62.6 (15 – 86) vs 45 (20 – 69)	Axial MRI	Axial AP canal diameter is shorter in cases than controls. Relative DLSS: Axial AP canal diameter of L1<20mm, L2<19mm, L3<19mm, L4<17mm, L5<16mm, S1<14mm. Critical stenosis: L4 < 14mm, L5 < 12mm, S1 < 12mm.
Chatha *et al*[16], 2011	Retrospective cohort study	Moderate	100 cases of possible metastatic disease without secondary spinal tumours (36 M, 64 F); 61.9 (4 – 94)	Midsagittal MRI	Canal Diameter was narrowest at L5-S1 (mean = 11.6mm) and widest at L1-L2 (mean = 15.6mm). A cut-off limit of the sagittal AP vertebral canal diameter for DLSS was proposed as 9mm at the vertebral body and disc level.
Kitab *et al*[2,17], 2018	Prospective cohort study	Moderate	709 LSS patients (306 M, 403 F); 50.8 (16 – 82)	Axial and sagittal MRI	No significant differences between lumbar canal dimensions and stenosis grades were found between the two cohorts in L1-L5 after adjusting for age, and there were no statistically significant variances in terms of global degenerative variables, except at L4-S1. Global degenerative variables included disc herniation, disc

					height, disc degeneration grade, endplate shape, Modic changes, Schmorl's nodes, facet degeneration grades, irregularities and sclerosis. Moreover, the authors found age-related degeneration in L1-L4 was more than in L4-S1.
Singh *et al*[18], 2005	Prospective cohort study	Low	15 DLSS surgical patients (13 M, 2 F) vs 15 age- and sex-matched controls (14 M, 1 F); 51.7 (43-65) vs 50.7 (41-55)	Axial and sagittal MRI	Cross-sectional area of spinal canal, pedicle length, axial AP vertebral canal diameter, vertebral body width, and sagittal AP vertebral canal diameter were found to be shorter in patients with DLSS ($P<0.05$). AP vertebral body diameter, canal width, pedicle width and sagittal vertebral body diameter and height were found to be statistically insignificant.
Kitab *et al*[5], 2013	Prospective cohort study	Low	66 DLSS patients (44 M, 22 F) vs 45 controls (31 M, 14 F); 40.7 (17 – 50) vs 39.5 (16 – 50)	Axial and midsagittal MRI, AP and lateral standing plain radiographs	Reduction in: 1. Spinal canal cross-sectional area to vertebral body cross-sectional area ratio on MRI ($p<0.001$). 2. AP spinal canal diameter to vertebral body diameter ratio on MRI ($p<0.01$) 3. Interpedicular distance to vertebral body diameter ratio on plain radiograph ($p<0.04$)

					4. Interlaminar angle (p<0.024) 5. Transverse spinal canal diameter to vertebral body diameter ratio (p<0.001)
Cheung *et al*[19], 2017	Case-control study	Low	66 DLSS patients (32 M, 34 F) vs 81 controls (31 M, 50 F); 65.9 (±SD 10.9) vs 56.4 (±SD 6.8)	Plain X-Ray	On AP radiographs, the axial vertebral body height and width and interpedicular distance were measured. On lateral radiographs, pedicle width (PW), sagittal vertebral body height and width (SBW), foraminal width, and posterior pedicle margin were measured. Sagittal vertebral body width:pedicle width ratio has the highest sensitivity (79%-92%) and specificity (50%-99%) to define DLSS under receiver operating characteristic analysis. Cut-off ratios are L1 > 2.0, L2 > 2.0, L3 > 2.2, L4 > 2.2, L5 > 2.5, S1 > 2.8
Mrowka *et al*[20], 1986	Case-series	Very low	29 DLSS patients with symptomatic sciatica (24 M, 5 F); N/A	Routing or tomographic X-rays, contrast examination	X-Rays failed to identify 2 cases of constrictions of spinal lateral recess. There was no correlation between narrowing of contrast column and radiological signs of narrowing

Postacchini *et al*[6], 1981	Case-series	Very low	8 DLSS patients diagnosed with water-soluble myelography	water-soluble myelography followed by CT	They found no relationship between AP vertebral canal diameter at the vertebral body level or severity of the laminal-facetal abnormalities with CT and the sites of myelographic changes.
Postacchini *et al*[7], 1980	Cross-sectional study	Low	8 DLSS patients vs 21 controls (11 M, 10 F); (24 – 42)	CT	Most cases had the shortest canal diameter at L4-L5. Multi-level stenosis and reduced laminae length were also observed in all cases, whereas interpedicular diameter and size of bony canal to size of vertebral body ratio varied.
Akar *et al*[21], 2019	Retrospective cohort study	Low	48 DLSS patients (21 M, 27 F) vs 52 degenerative LSS patients (26 M, 26 F); 58.8 vs 56.5	CT	Pedicle length was the only imaging parameter that was significantly shorter in the DLSS group (P=0.002), while facet joint angles, facet tropism degrees, lateral recess height and ligamentum flavum thickness appeared to be similar and statistically insignificant between groups (P=0.15-0.87).

DLSS indicates developmental lumbar spinal stenosis; LSS, lumbar spinal stenosis; MRI, magnetic resonance imaging; AP, anteroposterior; CT, computed tomography.

Table 4. Studies that Examined DLSS Treatment with Surgical Interventions and Their Outcomes.

Study	Design	Level of Evidence; Quality Score	Sample Size	Sex; Mean Age (Range)	Average Time of Follow-up (Range)	Methods of Diagnosing DLSS	Indications for Operation
Reale *et al*[9], 1978	Prospective cohort study	Low	37 patients with DLSS vs 95 patients with degenerative lumbar spinal stenosis	DLSS: 27 M, 10 F; 52.5 Degenerative: N/A	19 months	Lumbar myelography with low dose water-soluble contrast medium: AP view showed partial block; Lateral view showed narrowed dural sac etc. Definitive diagnosis made during operation.	N/A
Louie *et al*[24], 2017	Retrospective cohort study	Low	26 patients with DLSS vs 144 degenerative lumbar spinal stenosis	N/A; 47.1 vs 66.7	27.6 months	Plain radiographs revealed abnormally short pedicles and reduced cross-sectional area of the lumbar spinal canal.	Patients failed conservative treatments with symptomatic radiculopathy and neurogenic claudication.
Lee *et al*[26], 1978	Prospective cohort study	Very low	16 patients with DLSS and only 10 underwent	N/A	N/A	Myelographic examination showing concentric stenosis, sagittal flattening, or abnormal articular processes.	1. Intolerable pain even with supportive treatment;

			surgical intervention				2. Progressive muscle weakness; 3. Sphincter dysfunction.
Dai *et al*[25], 1996	Prospective cohort study	Very low	42 patients with DLSS	29 M, 13 F; 31.7 (19 – 44)	4.4 years (2 – 7 years)	Lateral lumbar radiographs: Below 15mm; Myelography: Narrowing or obstruction of contrast; CT: AP diameter less than 15mm, trefoil shape canal.	N/A
Verbiest[10], 1977	Case-series	Low	92 patients with DLSS	N/A	(1 – 20 years)	Relative stenosis: Mid-sagittal canal diameter of less than 12mm. Critical stenosis: Mid-sagittal canal diameter of less than 10mm. Mixed stenosis: Mid-sagittal canal diameter between 10mm and 12mm.	N/A
Cheung *et al*[8], 2019	Retrospective cohort study	Moderate	235 patients with DLSS	129 M, 106 F; 66.8 (± 11.3 s.d.)	10.1 years (± 4.8 s.d.)	MRI AP canal diameter: L1 < 20mm, L2 < 19mm, L3 < 19mm, L4 < 14mm, L5 < 14mm, S1 < 12mm.	Matching clinical symptoms with radiological findings of spinal levels that required decompression

Table 4. *(continued)*

Choice of Surgical Interventions	Assessment of Outcomes	Complications
For patients with DLSS, laminectomy of the whole segment supplemented by medial or complete removal of articular facets were conducted. For patients with degenerative LSS, extended laminectomy with bilateral foraminotomy and medial or complete facetectomy, or interhemilaminectomy were used.	Divided into groups: Excellent (back to work and free/nearly free from pain); good (back to work with recurring pain); poor (others); unknown (loss of follow-up). Preoperative and postoperative symptoms were recorded.	5 DLSS patients underwent reoperation due to wound infection, epiduritis, and spondylitis. Preoperative symptoms were worsen in some cases.
All patients had laminectomy with a Kerrison rongeur at the symptomatic level by using the standard posterior approach. Spinous processes were only removed at the necessary levels. Laminae were thinned with a rongeur and high-powered burr. Medial facetectomy and foraminotomy were performed to ensure adequate decompression. Extruded discs were also removed in several cases.	Comparison between preoperative and postoperative visual analogue scale and Oswestry Disability Index. Postoperative complications were also assessed.	Complications including dural tear, recurrent symptomatic pain at the back and lower extremities and reoperation were found to be statistically insignificant between groups (P=0.089-0.719).
Concentric stenotic patients had resection of entire articular process and bilateral laminae; sagittal flattening patients had total removal of laminae; abnormal articular	Scoring system was implemented, assessing pain sensation, sitting endurance, walking distance, night pain, ambulation, sphincter function, ability to lift, and	N/A

process patients had resection of the abnormal articular process and ipsilateral laminae.	muscle witness. They were added up to a total of 100 points. < 30 points: Poor 30 – 50 points: Unsatisfactory 50 – 75 points: Satisfactory 75 – 100 points: Excellent	
Simple discectomy was performed through laminotomy at one level (L4-L5 or L5-S1). Inferior margin of laminae and medial inferior facet were removed first, followed by ligamentum flavum. Some cases further required superior marginal laminotomy.	Evaluation system was implemented by comparing preoperative and postoperative symptoms. Divided into excellent, good, fair, or poor	No complications
Chisel and mallet for removal of thickened laminae, unroofing the intervertebral foramina with a chisel by removing the inferior articular process first, and starting laminectomy from the next normal space in an interlaminar space obliterated by overlapping laminae.	Preoperative symptoms and postoperative symptoms were compared, including intermittent claudication, lumbago, sciatica, neural deficit.	Radicular deficit, vertebral displacement, post-operative ossifying arachnoiditis, annular non-ossifying arachnoiditis, recurrent stenosis, and reoperation.
Bilateral fenestration by laminotomy with undercutting of cranial lamina and laminotomy of caudal lamina until ligamentum flavum was detached. Then, medial	N/A	N/A

facetectomy of articular processes and removal of ligamentum flavum.		
DLSS indicates developmental lumbar spinal stenosis; AP, anteroposterior; CT, computed tomography; MRI, magnetic resonance imaging.		