Summary

Background: Multiple reports of congenitally absent long head of biceps tendon (LHBT) have been reported in the literature. However, there is no consensus on the clinical implications of this relatively rare entity.

Study Purpose: To systematically review and analyze all studies which have reported absence of LHBT.

Methods: PubMed and EMBASE databases were searched. Also, a secondary search was performed by pearling the bibliography of all the full-text articles obtained. Pre defined inclusion criteria was used for abstract screening by two independent observers. Twenty three studies met our inclusion criteria, were included for the final analysis and the data was pooled. The cases were further sub-grouped according to the classification of Dierickx et al.

Results: Till date, 35 cases of absent LHBT have been reported. Males and females were equally affected. Eight of these were bilateral and only four cases had other associated congenital anomalies. Majority of the patients presented with shoulder pain (85.7%) while 37.1% had shoulder instability (mainly anterior instability). The ABS type was the most common variant reported. The finding was missed in 60% of the cases on the initial MRI only to be detected later on shoulder arthroscopy.

Conclusions: Congenitally absent LHBT may not be as rare as was previously thought to be. Due to the heterogeneity and the low level of evidence of the data available, it is hard to conclude if a congenitally absent LHBT is a cause of shoulder pain/impingement or instability on its own.

Level of evidence: IV.

KEY WORDS: arthroscopy, shoulder, long head biceps tendon, congenital variations.

Introduction

Congenital variations of the long head of biceps tendon (LHBT) have hitherto been thought to be extremely rare1-4. However with the advent of magnetic resonance imaging (MRI) and arthroscopy, the rate of detection and diversity of congenital variants of LHBT has increased significantly5. Absent LHBT is one such variant and has been documented in many cadaveric, imaging and arthroscopic studies5,6. Clinical diagnosis is extremely challenging and somewhat impossible due to the rarity of the condition, lack of awareness, variable clinical presentation, a lack of specific clinical signs and limitations of the conventional MRI7. Shoulder arthroscopy is the gold standard modality for diagnosis; hallmark is the absence of the intra-articular portion of LHBT in the presence of a shallow or absent bicipital groove3,6. A lack of history of significant trauma, absent Popeye’s sign on clinical examination and supportive MRI/MRA findings differentiate this anomaly from a biceps tendon rupture4,6,8.

Dierickx6 presented a comprehensive classification of congenital anatomic variants of the LHBT in 2009. Three possible pathoanatomical variations in this classification could present with absence of the LHBT on shoulder arthroscopy and MRI – these are ABS, ADH-CL and ADH-CO types. The ABS type is characterized by the complete absence of the intra-articular and extra-articular parts of the LHBT. In the ADH-CL category, the LHBT inserts normally on the labrum or supraglenoid tubercle but is adherent to the undersurface of cuff (i.e. the tendon is fixed at one end). In the ADH-CO type, the LHBT merges with the cuff completely with no fixed ends (Fig. 1)6.

It is not clear if different congenital variations which can lead to arthroscopic absence of LHBT have the same presentation6. In this systematic review, we have pooled data from all studies which have reported arthroscopic absence of LHBT in the literature so far. The cases have been classified according to the Dierickx6 classification after analyzing the clinico-radi-
ologic details provided by the Authors into three subtypes (ABS, ADH-CL and ADH-CO) to facilitate further analysis.

Methods
Ethical committee review was not necessary for this study as it is a systematic review of the literature. All research was carried according to the ethical guidelines followed by this Journal\(^9\). Standard PRISMA guidelines and checklist was used for the construction of this study\(^10\). The review was registered in the PROSPERO database prospectively and the protocol is available online (study registration number: CRD42017058939).

Search Methodology
PubMed and EMBASE databases were searched from their period of inception up to 7\(^{th}\) May 2017 using the keywords as shown in Table I. Non-English articles were exempted from the review. Additionally, a secondary search or pearling was performed by manually screening the bibliography of all selected full-text articles for additional reports.

Inclusion and exclusion criteria
We included clinical studies of any design if the primary research question of the study was related to the absence of LHBT. Cadaveric studies, narrative reviews and conference abstracts were excluded from the review and so were non-English articles.

Figure 1. An illustration depicting the subtypes of LHBT anomalies that might give an impression of arthroscopic absence of the tendon according to the Dierickx et al. classification: A) Normal joint; B) ABS type-complete absence of LHBT; C) ADH-CO: Complete adherence of the tendon to the undersurface of the rotator cuff; D) ADH-CL: LHBT inserts normally on the labrum or supraglenoid tubercle but is adherent to the undersurface of cuff.
The study results were screened and analyzed independently by 2 reviewers (R.J. and D.K.C). The study title was utilized to screen for potentially eligible studies, and the abstract of all selected studies was analyzed in detail to determine inclusion. In cases of uncertainty, inclusion or exclusion was ascertained after going through the full-text of the article in question. Full-text was obtained for all studies that were included for the final review. Any conflict was resolved by further review, discussion and consensus between the two reviewers.

Data from the included studies was collected on structured abstract forms. This included Author name and year of publication, demographic features, clinical presentation details, radiologic findings and details of management. The arthroscopic pathoanatomic details of the intra-articular portion of the LHBT were analysed and grouped as per the classification given by the Dierickx et al. for further analysis. This data was summarised in a tabular format (Tab. II).

### Results

The title word search identified 899 studies out of which a total of 23 studies were included for the final analysis. Nineteen of these 23 studies were case reports and four were case series. Till date, 35 cases have reported absence of LHBT. These studies have been published between 1997 and 2016 (Tab. II).

#### Clinical presentation

Gender ratio of cases was 1:1 (17 females and 17 males; in one case, gender wasn’t mentioned). A bilateral presentation was observed in 8 cases and a unilateral presentation in 27 cases (right shoulder in 12 cases, left shoulder in 7 cases, 8 bilateral, side not mentioned in 8 cases). The affected shoulder was the dominant shoulder in 8 patients, non-dominant shoulder in 3 patients (dominance of the affected shoulder was not mentioned in 16 cases). Overall, 30 out of the 35 cases presented with shoulder pain and 13 cases had symptoms of shoulder instability (NA in 6 cases). Among these 13 cases, 12 had signs and symptoms of anterior instability while one patient had posterior instability associated with glenoid dysplasia. Nine patients had positive outlet impingement signs on physical examination (details not specified in 7 cases). Only 2 patients had associated generalized ligament laxity. Four patients had other associated congenital anomalies- VATER, radial ray, upper limb anomalies, spina bifida occulta, congenital inguinal hernia and undescended testicle are the disorders which have been noted.

#### MRI & arthroscopy findings

On MRI, a rotator cuff pathology was observed in 36% cases (9/25) compared to labral pathology which was observed in 40% cases (10/25 cases). On arthroscopy, additional labral pathology was detected in five cases whereas one case had an additional infraspinatus tear.

#### Arthroscopic Classification (according to Dierickx et al.)

When sub-classified according to Dierickx et al. classification, the incidence of ABS type was 39.4% (most common type with 13/33 cases). ADH-CL was 36.4% (12 cases) and ADH-CO was 21.2% (7 cases). We could not apply this classification in three cases.
Table II. Search strategy used for the systematic review in PubMed and EMBASE databases.

<table>
<thead>
<tr>
<th>S.No</th>
<th>Author (Year)</th>
<th>Case(s)</th>
<th>Age/Sex/Occupation</th>
<th>Side/Dominance</th>
<th>Clinical Presentation</th>
<th>Onset</th>
<th>Radiologic Findings</th>
<th>Additional Arthroscopic Findings</th>
<th>Classification*</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Kim et al. 2009</td>
<td>1</td>
<td>52/F/</td>
<td>Rt/Dom</td>
<td>Pain, Weakness Impingement</td>
<td>-</td>
<td>MRI – SSP tear (Bursal side) Retrospective MRA confirmed abnormal course</td>
<td>Partial tear of ISP</td>
<td>ADH-CL</td>
<td>Rotator cuff repair (ISP) Nothing for LHBT</td>
</tr>
<tr>
<td>2</td>
<td>MacDonald 1998</td>
<td>1</td>
<td>25/M/</td>
<td>Labourer</td>
<td>LV-</td>
<td>Pain Impingement</td>
<td>Insidious</td>
<td>No MRI details given</td>
<td>Slight fraying of superior labrum LHBT completely adherent to cuff</td>
<td>ADH-CO</td>
</tr>
<tr>
<td>4</td>
<td>Egea et al. 2010</td>
<td>1</td>
<td>43/M</td>
<td>Rt/Dom/</td>
<td>Pain</td>
<td>Acute</td>
<td>MRI-1 – Antero-inferior labral tear, bursitis and SSP tendinitis MRI-2 – Bursitis, SSP tear absent intra-articular part LHBT</td>
<td>SLAP tear, absent LHBT</td>
<td>Not defined</td>
<td>Surgery 1- Arthroscopic SLAP repair Surgery 2- SSP repair with SAD and damaged repaired SLAP Surgery 3- Open intervention; Biceps tendon merging with SSP – tenotomy, tenodesis &amp; SSP repair</td>
</tr>
<tr>
<td>5</td>
<td>Gaskin et al. 2007</td>
<td>2</td>
<td>45/M</td>
<td>LV/-/-</td>
<td>Pain</td>
<td>Pain, RDS</td>
<td>Chronic</td>
<td>Chronic</td>
<td>1. MRA – antero-inferior labral tear, superior labral tear, absent LHBT 2. MRA – abnormal course and hypoplastic LHBT</td>
<td>Anteroinferior labral injury, Retrospective MRA confirmed finding Anteroinferior labral tear</td>
</tr>
<tr>
<td>6</td>
<td>Hyman et al. 2001</td>
<td>1</td>
<td>17/M</td>
<td>+/- Dom/High school wrestler</td>
<td>RDS</td>
<td>Acute</td>
<td>MRI – Attenuated capsule, Bankart and Hill Sachs lesion, unremarkable LHBT</td>
<td>Voluminous capsule</td>
<td>ADH-CL</td>
<td>Arthroscopy(open), Bankart repair &amp; superior capsuloraphy</td>
</tr>
<tr>
<td>7</td>
<td>Gillardin et al. 2013</td>
<td>1</td>
<td>22/M</td>
<td>RV/-/-</td>
<td>Pain (Associated with VATER syndrome)</td>
<td>Acute</td>
<td>USG – Absent LHBT MRI – Same Findings</td>
<td>Conservatively treated</td>
<td>ABS</td>
<td>Conservatively treated</td>
</tr>
<tr>
<td>8</td>
<td>Costa et al. 2016</td>
<td>1</td>
<td>29/M</td>
<td>RV/Fighter</td>
<td>Pain</td>
<td>Insidious/chronic</td>
<td>USG – Bilaterally absent LHBT MRI – Same findings plus shallow intertuberular sulci</td>
<td>Details not known</td>
<td>ABS (Bilateral)</td>
<td>Details not known</td>
</tr>
<tr>
<td>9</td>
<td>Foad et al.</td>
<td>1</td>
<td>24/F</td>
<td>LV/Dom/Chiropractor</td>
<td>Pain</td>
<td>Instability</td>
<td>Chronic</td>
<td>MRA – LHBT medial subluxation suspected</td>
<td>Anteroinferior labral tear</td>
<td>ABS</td>
</tr>
<tr>
<td>10</td>
<td>Franco et al. 2005</td>
<td>1</td>
<td>37/M</td>
<td>Bilateral</td>
<td>Pain, weakness (bilateral) Generalized ligament laxity Associated with spina bifida occulta, congenital inguinal hernia &amp; undescended testicle</td>
<td>Chronic</td>
<td>Lt side- Labral tear, diminished LHBT Rt side- Labral tear</td>
<td>Lt side- Bankart lesion, hypoplastic LHBT Rt side – Absent LHBT</td>
<td>ABS</td>
<td>Bankart repair, capsular plactation (bilateral)</td>
</tr>
<tr>
<td>No.</td>
<td>Author et al.</td>
<td>Gender</td>
<td>Side</td>
<td>Pain</td>
<td>Instability/Disability</td>
<td>Imaging</td>
<td>Procedure</td>
<td>Additional Details</td>
<td></td>
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<tr>
<td>11</td>
<td>Glueck et al. 2003</td>
<td>1</td>
<td>25/F</td>
<td>Bilateral</td>
<td>Pain (Bilateral) Instability (bilateral) Generalised joint laxity</td>
<td>Chronic</td>
<td>Normal MRI pre-operatively Absent LHBT noted retrospectively</td>
<td>Capacious capsule with positive drive-through sign</td>
<td></td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>Koplas et al. 2009</td>
<td>1</td>
<td>40/M</td>
<td>Bilateral</td>
<td>Pain (Bilateral) Insidious/chronic</td>
<td>MRA – Absent LHBT with SSP and superior labral tear (Bilaterally) with smooth shallow intertubercular sulci</td>
<td>MRA findings confirmed on arthroscopy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>Kuhn et al. 2009</td>
<td>1</td>
<td>30/M</td>
<td>Bilateral/radiology technician</td>
<td>Pain (Bilateral) Insidious/chronic</td>
<td>MRA 1 (Rt side) – “Rupture” LHBT, SSP and SLAP tear, MRA 2 (Lt side) – Absent LHBT, SSP and SSP partial tears</td>
<td>Buford complex</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>Maldjian et al. 2014</td>
<td>1</td>
<td>42/M</td>
<td>Bilateral</td>
<td>Pain</td>
<td>Acute (Post trauma) Association with congenital radial ray deformity</td>
<td>MRI – Absent LHBT, Labral tear, Bilateral shallow bicipital groove</td>
<td>MRI findings confirmed on arthroscopy Prominent rotator cable noted</td>
<td></td>
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</tr>
<tr>
<td>15</td>
<td>Mariani et al. 1997</td>
<td>1</td>
<td>23/M</td>
<td>Rt/-Policeman</td>
<td>Pain</td>
<td>Acute (Post trauma)</td>
<td>MRI – Increased signal intensity SSP tendon Post operative bilateral MRI- Hypoplastic LHBT No other abnormalities detected Not classifiable Diagnostic arthroscopy (Rt side)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>Dierickx et al. 2009</td>
<td>2</td>
<td>35/M 42/F</td>
<td>Pain Instability</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>ABS No LHBT specific procedures in ABS cases Bursectomy &amp; capsular reconstruction</td>
<td></td>
<td></td>
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<tr>
<td>19</td>
<td>Dierickx et al. 2009</td>
<td>1</td>
<td>62/F</td>
<td>Pain</td>
<td>-</td>
<td>SSP tear (Full thickness)</td>
<td>ADH-CO Tenodesis LHBT with interf screw Mini-open SSP suture</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>21</td>
<td>Audenaert et al. 2008</td>
<td>1</td>
<td>36/M</td>
<td>Rt/Dom/-</td>
<td>Pain</td>
<td>Chronic/Insidious No details of MRI Plain radiograph- Degenerative arthritis of RI AC joint</td>
<td>The long head of the biceps was visible as an extrasynovial impression running from the supraglenoid tubercle to the intertubercular groove</td>
<td>ADH-CL SAD &amp; Distal clavicle resection</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
due to the lack of adequate details\textsuperscript{1,22,24}. It is to be noted that six out of the eight cases with a bilateral presentation were of the ABS type; one case was ADH-CO type and another was not classifiable\textsuperscript{2,3,5,13,16,19,20,22}.

**Discussion**

Initially thought to be a vestigial structure of no functional significance by Lipmann et al.\textsuperscript{26}, the LHBT has received tremendous attention regarding its anatomy and clinical function in the recent past. The long head of the biceps tendon has now been described as a dynamic stabilizer of the shoulder joint as it acts as a depressor of the humeral head and also as an elevator of the superior glenoid labrum\textsuperscript{27,28}. This has been proven on the basis of different cadaveric and electromyographic studies\textsuperscript{29,30}.

The first anatomic description of congenital variations of the LHBT was described by DePalma in his textbook in 1983\textsuperscript{31}. Mariani et al. in 1997 published the first case report documenting arthroscopic absence of LHBT in a patient without traumatic history\textsuperscript{22}. Since then, many case reports and series have reported cases where the LHBT was absent. Different embryological hypothesis have been put forward to explain the congenital absence or deficiency of the LHBT\textsuperscript{5,6,13}. Whatever is the embryological origin of these variations, the clinical presentation and pathoanatomy in these cases has been highly variable as reported above.

Ghayalini et al. initially classified congenital absence or deficiency of the LHBT into 4 types\textsuperscript{5}. In type 1 lesions, the tendon was completely absent and so was the supraglenoid tubercle. In type 2, a remnant of the LHBT is present but the supraglenoid tubercle is still absent. In type 3 lesions, both the supraglenoid tubercle and LHBT is present; however, the LHBT is embedded in the capsule. In type 4, the tendon is present but hypoplastic\textsuperscript{5}.

Dierickx et al.\textsuperscript{6} have published the largest case-series of congenital variations of LHBT till date in their multicenter collaboration. Out of a total of 3000 shoulder arthroscopies (1500 each in Belgian and Italian populations respectively) conducted over a period of ten years (1996-2006), they identified 57 shoulders with congenital anomalies. They formulated an all-inclusive classification dividing these congenital anomalies into four major families with each subtype further divided into four or five subgroups. The four major families are- MESO (mesotenon family), ADH (adherent LHBT), SPL (split LHBT) and ABS (absent LHBT or agenesis). It is to be noted that both ABS and two subgroups of the ADH type (ADH-CL and ADH-CO) can give an impression of absence of the LHBT on arthroscopy and MRI. The incidence of ABS, ADH-CL and ADH-CO variations among the different congenital LHBT anomalies are 3.5, 8.8 and 1.7% respectively\textsuperscript{6}.

The ADH-CL and ADH-CO subtypes are the cases with abnormal origin and intra-articular course of the LHBT,
where the tendon adherent to the superior capsule or the under surface of the supraspinatus muscle (equivalent to Ghayalini type 3 variant). It has been hypothesized that due to the adherent intra-articular course, the gliding of the biceps tendon is hampered which tends to pull the rotator cuff downwards leading to impingement and eventually a rotator cuff tear6.

It is tough to diagnose this condition on the pre-operative MRI unless the radiologist keeps a high index of suspicion. This is the reason why 60% of the cases were missed on the initial MRI. Also, a MR arthrogram is superior to the conventional MRI in diagnosing the absent LHBT4,32. Most cases have been diagnosed incidentally during shoulder arthroscopy followed by a retrospective confirmation on the initial MRI or a fresh MR arthrogram.

In most of the cases, no active surgical intervention was performed for the absent or adherent tendon and it was just recorded as an incidental finding. A tenolysis/marsupialisation procedure has been performed in three cases and a tenodesis has been performed in one case6,6. Open exploration of the tendon was conducted in one case12. The functional outcomes do not vary between the cases where a tenolysis/tenodesis was performed and in those cases where the anatomical variant was left undisturbed.

It is clear from our review that the clinical picture of the patient at the time of presentation can be highly variable; they can present with pain, instability, impingement with rotator cuff and/or labral pathology. It is not clear if an absent LHBT is just an incidental finding or a predisposing factor to shoulder pain and/or instability. Lack of data on incidence of absent LHBT in asymptomatic shoulders compounds the problem. Due to the low level of evidence available (all articles are case reports/series) and the non-uniformity of the data in these case reports, further statistical analysis is not feasible and has high chances of being inaccurate.

Conclusions

To summarize, not all cases with absence of LHBT are the same, and different pathoanatomical variants can exist; this needs careful appraisal and understanding. The clinical picture at presentation is highly varied; an association between the pathoanatomy of the congenitally absent LHBT and the clinical presentation could not be established by our review. The clinical relevance lies in the fact there is no significant evidence that an absent LHBT acts as a predisposing factor for rotator cuff or labral pathology; this cannot be ascertained with the current available evidence, and it is possible that an absent LHBT may just be an incidental finding of no clinical significance.

References