Cleft hand classification and treatment: literature review

Classificação e tratamento da mão em fenda: revisão da literatura

Cleft hand is a rare congenital deformity characterized by a longitudinal deficiency of the central rays of the hand, which may be associated with other malformations. Due to the wide spectrum of manifestations, the treatment is challenging. This study presents the most suitable classifications, surgical techniques, and follow-up adopted according to the clinical manifestation. The search was performed in the Web of Science, PubMed, Scopus, Cochrane and Embase databases, descriptors and terms related to the hand anomaly in a typical cleft. Thirty-two articles were included and analyzed regarding the classification of the anomaly, classification of the severity of expression, surgical techniques and studies adopted with information on the surgical intervention for a cohort of patients. Considering that studies about cleft hands could be directly affected by embryological, genetic and molecular biology discoveries, different classifications have been described, and several studies to complement existing surgical techniques have been found. Innovative studies are scarce. In addition to better quality research, standardization in the description of techniques and results could elucidate existing treatment options gaps.

Keywords: Congenital abnormalities; Hand; Finger joint; Congenital, Hereditary, and neonatal diseases and abnormalities; Hand deformities.

INTRODUCTION

Congenital cleft hand (CCH) was originally classified as typical versus atypical cleft hand. With the advancement of genetics and molecular biology, the atypical cleft hand was reclassified as a teratological sequence of symbrachydactyly.

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This anomaly is characterized by the “V” shape, which may be associated with the absence of one or more digits, and may be unilateral or bilateral, with or without the involvement of the feet. Generally, it is an autosomal dominant inheritance, with variable penetrance and expressiveness.

Resulting from a longitudinal deficiency of the central rays of the hand, CCH can range from a simple skin cleft of soft tissues to the suppression of all rays except the smallest digit. Based on the three axes of hand and upper limb development, CCH is currently classified as hand plate malformations - abnormal axis differentiation (patterning/late limb differentiation).

Manske & Halikis and Sharma and Sharma stand out among the most used classifications. The first is based on the involvement of the first commissure. The second provides a complete hand description and assigns a numerical value to each element, with the subsequent recommendation of the indicated surgical procedure.

Indications for surgical treatment range from space deficiency in the first commissure, absence of the thumb, progressive deformity to severe flexion contractures of one or more fingers. However, this topic remains controversial and challenging, especially due to the patient’s adaptation to the deformity and acceptable functionality of the limb.

**OBJECTIVE**

This literature review aims to present the classifications, the most relevant surgical techniques reported in the literature and the results obtained from the studies included.

**METHODS**

**Databases and research**

The bibliographic search was carried out between April and October 2020 in journals indexed in the Web of Science, PubMed, Scopus, Cochrane and Embase databases. The search terms used were a combination of “Typical Cleft Hand,” “Cleft Hand,” “Ectrodactyly,” “Central hand,” “Central ray deficiency,” “EEC syndrome,” “Cleft hand,” “Cleft-hand,” “Cleft-Hand Malformation,” “Lobster claw,” “Fingers/abnormalities,” “Muscle, Skeletal/abnormalities,” “Hand Deformities, Congenital/pathology,” “Trigger Finger Disorder/congenital,” “SHFM”, “Collateral Ligaments/surgery”, “Hand Deformities, Congenital/ surgery”, “Metacarpophalangeal Joint/surgery”, “Tendon Transfer”, “Surgical Flaps”, “Suture Techniques”, “Syndactyly/surgery”, “Reconstructive Surgical Procedures”, “Hand deformities, Congenital/surgery”, “Fingers/surgery”, “Congenital/surgery”, “Treatment”, and other related terms. All records returned by the search were imported into Mendeley’s bibliographic management software, and duplicate publications were removed. We also identified relevant articles through bibliographic linking with eligible articles.

**Selection of studies**

The included studies were related to CCH and may have the following approaches: classification of the anomaly, classification of severity of expression, surgical techniques, intervention or cohort of patients undergoing surgical treatment. The search did not limit language or study design. For intervention analysis, in order to observe current practical trends, the search was restricted to studies published between 2000 and 2020.

Studies that analyzed patients with cleft hands resulting from trauma sequelae or syndromic association, review articles or secondary analyses and publications that were incomplete or did not provide sufficient data for one of the outcomes of interest were excluded. Patient cohort data from studies approaching surgical techniques were not used for interventional analyses.

**Data extraction**

Two independent investigators reviewed the search results to select eligible studies using pre-established inclusion and exclusion criteria. Disagreeing decisions were discussed with a third reviewer. Data were extracted using a form according to predefined variables for each analysis. In order to summarize the findings in the literature, we chose to include a topic unifying surgical techniques and classification of severity of expression according to suggestive reports observed in the literature.

**RESULTS**

**Selection of studies**

Five hundred twenty-seven studies were identified by searching the descriptors in the databases. Three hundred sixty-nine articles were excluded due to duplicity or by title, abstract and/or keywords. After applying the determined inclusion and exclusion criteria, 36 were considered potentially eligible; four were discarded based on clinical outcome, lack of data or inadequate study design, resulting in 32 studies for analysis of results. In Figure 1, the flowchart of the search for studies in the chosen databases is represented.
Analyses of included studies

The included studies were divided into four groups: (1) congenital anomaly classification ($n=8$); (2) expression severity rating ($n=10$); (3) surgical techniques ($n=12$); and (4) intervention analysis ($n=4$). Of these, only two studies met the inclusion criteria in two concomitant groups; therefore, they were counted as unique inclusions11,12. Therefore, the main features of the 32 included studies stratified into groups are presented in subsequent topics.

CCH functional classification

The classification system for congenital limb malformations was developed by Swanson et al.13, based on the grouping of anomalies according to the part affected during development. This system, accepted by the American Society for Surgery of the Hand (ASSH), the International Federation of Societies for Surgery of the Hand (IFSSH) and the International Society for Prosthetics and Orthotics (ISPO), is called the IFSSH classification. Subsequently, the Japanese Society for Surgery of the Hand (JSSH) suggested modifications to include two groups: “Abnormal lightning induction” and “Unclassifiable cases”15,16. A new classification for congenital anomalies of the upper extremity, known as the OMT classification and considered an alternative to the Swanson, Barsky and Entin classification17,18, was presented by Oberg et al.19 in 2010. Since its publication, the OMT classification has been vigorously evaluated by several authors for its usefulness and reliability, and recently, in 2020, an update was published6.
Therefore, according to the current classification, CCH is classified as IB1IV: I- Malformation; B- Hand plate: abnormal axis differentiation (late limb standardization/differentiation); 1- Proximaldistal Axis; IV: Cleft hand (cleft foot/hand malformation)⁶.

Congenital differences can also be classified according to their severity of expression, which can help in the functional determination and treatment orientation. Due to the unpredictability and peculiarity of the phenotypic presentations of this anomaly, a large number of classification systems have been proposed, which may be based on the number of defective rays¹¹,²⁰,²¹, teratological mechanism of aplasia and synostosis²², first commissure contracture⁷,¹², the complexity of associated anomalies²³ and radiological morphology and cleft position - medioulnar and ulnar²⁴.

More current classifications tend to present a greater complexity of information. Valenti et al.²⁵ proposed a classification based on six groups, each with a therapeutic strategy based on describing all clinical and radiographic abnormalities observed. In line with this, Sharma & Sharma⁸ described a new comprehensive functional classification considering all morphological determinants of the anomaly, such as absent digits, associated anomalies, cleft location and thumb functional status, calling it DAST⁸.

Among these, the most widely used clinical classification is that of Manske & Halikis⁷, which is based on the condition of the first commissure, with type I being normal (normal web); type II (narrow web) with moderate (IIA) or severe (IIB) narrowing; type III (syndactylized web) fused first commissure, syndactyly between thumb and forefinger; type IV (merged web) the first commissure included in the cleft, there is no index finger and the thumb is unstable; and type V (absent web) with the absence of the commissure due to the absence of the thumb.

As a general rule, a sum of scores greater than 4 or individual scores in any morphological determinant of the anomaly greater than 2 indicates a potentially more complex deformity with less possibility of satisfactory functional and aesthetic results⁸.

**Surgical technique**

Recommended surgical procedures for treating central deficiency include cleft closure, reduction of the intermetacarpal space, the release of syndactyly, and excision of polydactyl or transverse bone elements when present. Different techniques can reduce intermetacarpal space and interventions secondary to cleft closure, and the most frequently reported in the literature, along with indications, advantages and disadvantages, as reported by the study when available, are listed in Figure 2.

**Surgical technique according to expression severity classification**

Grading systems are essential to facilitate communication and guide surgical reconstruction. Therefore, we present the behaviors most commonly reported by the included studies, stratified according to the classification by Manske & Halikis⁷.

In order to expose the advances in knowledge of this anomaly, we present in Figure 3 the Manske & Halikis⁷ classification and the recommendations for procedures suggested by some authors of the new classification system for hands with typical clefts using the DAST⁸ classification.

**Intervention analysis**

Surgical information from cohort studies of patients undergoing procedures for the treatment of cleft hands is presented in Chart 1.
Figure 2. Recommended treatment according to classification.

<table>
<thead>
<tr>
<th>Type I</th>
<th>Treatment according to Manske and Halikis⁷</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type II A</td>
<td>Deepening of the 1st commissure with local flap (z-plasty) / skin graft; Reconstruction of the intermetacarpal ligament; Bone excision; Corrective osteotomy.</td>
</tr>
<tr>
<td>Type II B</td>
<td>Deepening of the 1st commissure with local dorsal flap; Transposition of the index radius; intermetacarpal ligament; Bone excision; Corrective osteotomy.</td>
</tr>
<tr>
<td>Type III</td>
<td>Deepening of the 1st commissure with local dorsal flap; Transposition of the index radius; Reconstruction of the intermetacarpal ligament; Bone excision; Corrective osteotomy; Release of Syndactyly; Skin graft if needed.</td>
</tr>
<tr>
<td>Type IV</td>
<td>Release of syndactyly; Skin graft if needed.</td>
</tr>
<tr>
<td>Type V</td>
<td>Finger-to-thumb transfer when possible.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Treatment according to DAST⁸ classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
</tr>
<tr>
<td>D:0, 1; A:*; S:1, T:0</td>
</tr>
<tr>
<td>Objective: Close the 1st commissure</td>
</tr>
<tr>
<td>Technique: Barsky; and Foucher, Lorea, Hovius, Pivato, Medina, 2006²⁶</td>
</tr>
</tbody>
</table>

*Associated anomalies may or may not be present and should be addressed by an ancillary procedure.

Figure 3. DAST classification for cleft hand.

Numerical scale for the number of missing fingers

- 0 – None
- 1 – A missing finger
- 2 – Two missing fingers
- 3 – Three missing fingers
- 4 – Four fingers missing with only one finger edge remaining

Numerical Scale for Associated Anomalies

- 0 – None
- 1 – Polidactyly
- 2 – Syndactyly
- 3 – Constriction rings syndrome/ Contractures/ Abnormal bone fusion
- 4 – Wide cleft with transverse bones in the cleft
- 5 – Presence of more than one associated anomaly

Numerical scale for crack location

- 0 – Normal / No functionally significant anomaly
- 1 – Contracted commissure without syndactyly
- 2 – Partial syndactyly
- 3 – Complete syndactyly
- 4 – Thumb hypoplasia (Blauth I-II)²²
- 5 – Floating thumb or thumb agenesis (Blauth IV-V)²²

Numerical scale for the functional state of the thumb

- 1 – Central
- 2 – Radial
- 3 – Ulnar
## Chart 1. Surgical techniques, advantages and disadvantages.

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Technique</th>
<th>Advantages disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Barsky, 1964&lt;sup&gt;1&lt;/sup&gt;</td>
<td>Uses a retial place, pedicle foursquare, in diamond shape to recreate the commissure after the cleft is narrowed. <strong>Indication:</strong> Cleft hand.</td>
<td><strong>Benefits:</strong> Aesthetic improvement. <strong>Disadvantages:</strong> Insufficient functional concern; without reconstruction of the first commissure.</td>
</tr>
<tr>
<td>Snow and Littler, 1967&lt;sup&gt;28&lt;/sup&gt;</td>
<td>The cleft is elevated like a palmar flap, with a small radial flap preserved by recreating the commissure; the first commissure space is freed, which may require splitting the dorsal interosseous and surrounding fascia; the second metacarpal is transposed and attached to the remainder of the third metacarpal base; Fixation is obtained with axial and transverse wires, and the palmar flap is transferred, recreating the new commissure between the second and fourth ray. <strong>Indication:</strong> Third metacarpal segment present.</td>
<td><strong>Benefits:</strong> Functional; Gain cosmetic. <strong>Disadvantages:</strong> Risk of distal flap necrosis due to its high length-to-base ratio; Traction of the adductor and dorsal interosseous muscles may cause some radial angulation in the local translocation, incompletely correcting the central cleft.</td>
</tr>
<tr>
<td>Miura and Komada, 1979&lt;sup&gt;27&lt;/sup&gt;</td>
<td>Index transposition in a central position and palmar and dorsal redesign as separate flaps to create the first commissure. Cleft incised from side to side. The index finger is raised in its neurovascular bundles and transposed by osteosynthesis with the third metacarpal or by angulation osteotomy. <strong>Indication:</strong> Cleft hand with an adducted thumb.</td>
<td><strong>Advantage:</strong> Small flaps of random transposition of the dorsal and palmar skin. <strong>Disadvantage:</strong> Incidence in necrosis distal and contracture secondary</td>
</tr>
<tr>
<td>Ueba, 1981&lt;sup&gt;28&lt;/sup&gt;</td>
<td>Use of transverse flaps from any edge of the cleft and transposition of the index digit; Reconstruction of the intermetacarpal ligament by a free tendon. <strong>Indication:</strong> Total absence of the third metacarpal.</td>
<td><strong>Advantage:</strong> Improved aesthetics without changing the function of the hand. <strong>Disadvantage</strong> aesthetic of transferring the palmar to dorsal skin and dorsal to palmar skin.</td>
</tr>
<tr>
<td>Buck-Gramcko, 1985&lt;sup&gt;29&lt;/sup&gt;</td>
<td>Cleft narrowing, syndactyly separation, crossbones removal, correction of joint flexion contractures, rotation or wedge osteotomies for axial deviations and ulnar translocation of the index digit. <strong>Indication:</strong> Deep intermetacarpalpal ligament reconstruction.</td>
<td><strong>Advantages:</strong> Cosmetically acceptable without translocation. <strong>Disadvantages:</strong> Inadequate correction of the central space.</td>
</tr>
<tr>
<td>Ogino, 1990&lt;sup&gt;11&lt;/sup&gt;</td>
<td>The index and ring fingers reconstruct the deep metacarpaltransverse ligament using flexor sheaths (part of the A1 or A2 pulleys). <strong>Indication:</strong> Total absence of the third metacarpal.</td>
<td><strong>Advantages:</strong> Possibility of spontaneous correction of the deformity in flexion of the ring finger. <strong>Disadvantages:</strong> -</td>
</tr>
<tr>
<td>Upton, 2004&lt;sup&gt;12&lt;/sup&gt;</td>
<td>Wide incision that extends from the ulnar side of the cleft around the malpositioned index digit to the thumb; may include index transposition, metacarpal and/or phalangeal osteotomies, joint releases, phalangeal osteotomies, adductor pollicis muscle preservation, first dorsal interosseous muscle release, syndactyly separation(s), and thumb duplication correction. <strong>Indication:</strong> Typical cleft hand.</td>
<td><strong>Advantage:</strong> Provides clear identification of all anatomical structures of the palm. <strong>Disadvantage:</strong> Grasp and precision maneuvers remain poor despite considerable functionality.</td>
</tr>
<tr>
<td>Foucher, Loréa, Hovius, Pivato, Medina, 2006&lt;sup&gt;20&lt;/sup&gt;</td>
<td>Translocation in the radial direction of the ulnar finger(s) by intracarpal osteotomy; When necessary, a synostosis metacarpal can be performed in the same procedure. <strong>Indication:</strong> Type IIA of the Manske &amp; Halikis classification&lt;sup&gt;7&lt;/sup&gt;.</td>
<td><strong>Advantage:</strong> No functional loss; Good alignment of the second metacarpal with the radius. <strong>Disadvantage:</strong> Mobility between hamate and capitate is physiologically limited in all biomechanical studies.</td>
</tr>
<tr>
<td>Oberlin, Korch, Belkheyar, Touam, MacQuillan, 2009&lt;sup&gt;31&lt;/sup&gt;</td>
<td>Reverse policing: The incision wraps around the second digit in the middle, extends over the dorsal edge of the cleft, and ends on the radial side of the third digit, where the second commissure space should be created. The index metacarpal is released (extraperiosteally) and translocated into the space of the absent third ray. After internal bone fixation, the flap, with its volar cutaneous pedicle, is transposed to reconstruct the first space of the mesh. <strong>Indication:</strong> Type II of the Manske &amp; Halikis classification&lt;sup&gt;7&lt;/sup&gt;.</td>
<td><strong>Advantages:</strong> Preservation of the dorsal venous network; no need for grafting; It does not harm the normal musculature of the thumb. <strong>Disadvantages:</strong> Possibility of ectopic bone deformation; Index finger misalignment; Divergence of the metacarpals if reconstruction of the transverse metacarpal ligament is insufficent.</td>
</tr>
</tbody>
</table>

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**Classification and treatment of the cleft hand**

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DISCUSSION

Many cases of CCH do not require surgical treatment. Therefore, the follow-up to be adopted demands delicate management when outlining the indications for intervention and the surgical plan must specifically address the unique abnormality of the patient. Since several authors report the psychological consequences of CCH caused by social stigmas, members of an interdisciplinary team may be requested to advise families with this condition.

The main indication for correcting the deformity is restoring function as close to normal and as early as possible, with the esthetic correction being a natural result of the intervention. As described in the literature, surgeons must assess the spacing of the first commissure, the presence or absence of metacarpals in the central rays, transverse bones, syndactyly that may be present and, especially, the vascular supply to the hand. If the cleft hand is due to vascular insufficiency, as in cases of associated syndromes, or if there are other concomitant deformities, a stepwise surgical correction is suggested.

It has been observed that understanding of the etiology of congenital limb anomalies is restricted and continues to progress, and we confirm this in the recent modification of cleft hand subcategorization in the OMT classification update published in 2020.

A large number of classifications according to the severity of CCH expression were found, with Manske & Halikis being the most frequently used, being cited in 100% of the studies included for analysis of surgical intervention. However, the most complete and current classification with recommendations for surgical procedures and prognosis is that of Sharma & Sharma.

It is important to highlight that some studies were redundant, describing modifications of the skin incision or refining techniques already consolidated, with the main objective of correcting the first commissure. If the first commissure is normal, so-called ‘simple cleft closure’ is suggested, with the reconstruction of soft tissue, ligaments, tendons and bones. The incision can be closed using different methods; the skin is usually removed from the lateral side of the adjacent digits to form a longitudinal scar.
Often, a flap from the ulnar half of the cleft is used to increase the depth of the first commissure, creating a wider, more functional space between the index finger and thumb. Authors emphasize that detecting the neurovascular pedicle and careful division of the cleft are important steps in identifying additional tendons. According to studies included in the intervention analyses, if the second and fourth digits remain divergent, an osteotomy of the base of one of the metacarpal bones can be considered. In cases of transverse bones, total or partial removal is suggested, leaving parts of it in continuity with the metacarpal joints to avoid damage to collateral ligaments, instability or stiffness\textsuperscript{40}. Reconstruction of the deep, transverse metacarpal ligament can be performed using flexor sheaths (part of the A1 or A2 pulleys) of the index and annular digits, dorsal base rotation graft\textsuperscript{11}, tendon/fascia grafts\textsuperscript{36}, or even absorbable stitches and K-wires.

For complex cases with syndactyly of the first commissure, options are presented according to the severity of the narrowing. If the syndactyly is proximal and loose, it can be released with an appropriate flap (z-plasty, combined z-plasty, or rotation-transposition flap) in association with cleft closure, as described.

For severe narrowing, attention should be paid to the distal bifurcation of the neurovascular bundle, not only between the thumb and index but also between the ring and little digit. If microvascular procedures cannot solve this problem, authors warn of the possibility of necrosis\textsuperscript{41,42}. Therefore, the transposition of the second ray to the third metacarpal bone (if any) may be indicated. In advanced cases of partial or complete aplasia, transfer of toes or bone graft covered with periosteum is suggested\textsuperscript{42}. Conservative treatment is reserved for the most severe cases, in which there is usually a great functional adaptation and in patients with severe neuropsychomotor developmental deficit, making adequate rehabilitation impossible\textsuperscript{43,44}.

It was observed that practical application studies and long-term follow-up are limited. Only four studies that presented details of surgical intervention for a cohort of patients undergoing cleft hand treatment were identified, as shown in Chart 2. Of these, the most frequent distribution of cleft hands was type II. No type IV or V patients were described, a similar observation described by Manske & Halikis\textsuperscript{7}, who reported that they are the rarest subtypes and are difficult to manage, a factor that would justify the absence of studies containing these subtypes.

In recent studies, the Snow procedure is often related to its limitations, such as low viability of the palmar flap, technical difficulties, and records of complications resulting from necrosis. However, Rider et al.\textsuperscript{45}, when studying the technique, observed a low flap necrosis rate, but the revision was necessary for one third of the patients. Despite this finding, there is a relevant use of the technique by Miura & Komada\textsuperscript{27}, which is justified in the simpler design and less risk of flap necrosis while producing similar functional and cosmetic results\textsuperscript{39}. A study with long-term results of the technique by Miura & Komada\textsuperscript{27} demonstrated excellent patient satisfaction in function and esthetics\textsuperscript{39}. The same occurrence was reported in the literature for interventions performed using the Upton technique\textsuperscript{12}.

In general, patients with a cleft hand due to vascular deformity are at high risk of skin loss and poor perfusion of the surgical site after surgery, especially if the procedure is not staged properly. In addition, finger stiffness remains the most common postoperative complaint, despite improving functional results\textsuperscript{34}. Because of this, we emphasize that the median cleft of the hand is a complex but rare malformation that requires individualized management based on the severity of expression.

\textbf{Chart 2.} Intervention analysis of articles published in indexed journals included in the systematic review.

<table>
<thead>
<tr>
<th>Study (Author, year)</th>
<th>Patients/hands (n)</th>
<th>Manske type and (n)</th>
<th>Correction of central deficiency</th>
<th>Additional procedures (n)</th>
<th>Component and transverse bone (n)</th>
<th>Observation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rider, Grindel, Tonkin, Wood, 2000\textsuperscript{45}</td>
<td>12/12</td>
<td>Type IIB: (-) Type III: (-)</td>
<td>Snow &amp; Littler\textsuperscript{26}</td>
<td>Osteotomy: 3 Osteotomy for delta phalanx: 2 Revision of the first commissure: 3 Revision of the syndactyly scar: 1 Religation of the cleft: 1 None: 2</td>
<td>Dorsal base rotation graft\textsuperscript{11}, bone suture or tendon graft: 8</td>
<td>There were no cases of graft necrosis, although two grafts showed ischemia at the edge; Four (36%) secondary revisions of the first commissure were performed.</td>
</tr>
</tbody>
</table>

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CONCLUSION

Studies on FCM are directly affected by discoveries in embryological, genetic and molecular biology. During the last few years, advances in these fields have led to restructuring the classification system and understanding different presentations. Regarding treatments, pioneering techniques include cleft closure and reconstruction of the first commissure. The main complications described were problems with necrosis of the distal flap and stiffness. Several studies on updating these techniques were found. In addition to better quality research, standardization in the description of techniques and results could elucidate existing treatment options gaps.

COLLABORATIONS

AGPCP Data analysis and/or interpretation, Statistical analysis, Data Collection, Conceptualization, Conception and design of the study, Resource Management, Project Management, Investigation, Methodology, Carrying out operations and/or experiments, Writing - Preparation of the original

MSM Data analysis and/or interpretation, Methodology, Writing - Preparation of the original, Writing - Review and Editing

JCN Final Manuscript Approval, Project Management, Writing - Review and Editing, Supervision, Preview
REFERENCES


