

Sister Cases of "Delta Phalanx" which was Difficult to Differentiate from Brachydactyly^{*}

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ABSTRACT

The authors in October 1975 encountered a case that visited their hospital with the chief complaint of deformity of both middle fingers. The case closely resembled brachyhyperphalangism, but no definite diagnosis could be obtained as a findings suggestive of deltaphalanx was present in some phalanges and some took the view that polydactylous change of the middle finger was suggested. However, because numerous deltaphalanges were found in both index and middle fingers in a younger sister bone five years later, both of the cases were diagnosed as belonging to the category of what hand surgeons of Japan maintain is cleft hand group. However, there are literature which describe cases like these two as typical cases of brachydactyly, and it was considered that study would have to be made in future whether brachydactyly and cleft hand group had some parts of their developmental mechanism in common or whether the concept of brachydactyly is as yet unestablished.

CASES

Case 1.

The case is a girl born on 5 July 1975 as the first child of a 27-year-old father and a 26-year-old mother. There was no abnormality in particular during pregnancy, and delivery was full-term and easy. The child weighed 2880 g at birth. Already at birth, overlapping of the ring with the middle finger was observed in both hands, and because this deformity gradually aggravated, the case visited this department for examination in the fifth month after birth.

The external appearance at examination is as shown in Fig. 1. The hands shown almost identical appearance. The middle finger is remarkably flexed ulnarly at the PIP joint, the DIP joint is slightly deviated radially, and the middle finger as a whole is short. Active motion, which was satisfactory at the MP joint, was not observed at the PIP or the DIP joint. It was a very characteristic finding that the ring finger marked overlapped with the middle finger in flexed position. Roentgenographic

views are shown in Fig. 2. Both middle fingers have four phalanges and present so-called hyperphalangism. Development of the interphalangeal joint was poor in both middle fingers. (Fig. 1, Fig. 2)

In making diagnosis of this case, there were two conflicting views. One view had it that it belongs to the category of brachydactyly and was homologous to type C (Bell, 1951³⁾) which present hyperphalangism of index and middle finger, and the other view held that it belonged to the category of so-called cleft hand group (Watarai, 1984⁴⁾) which latent polydactylous change in the middle finger, from the fact that the most proximal phalanx showed typical delta phalanx. With no conclusion being obtained in the final analysis, diagnosis was reserved. Roentgenographic views of a case diagnosed as typical brachyhyperphalangism are shown in Fig. 3, which are very interesting indeed when compared with Fig. 2. (Fig. 3)

Case 2.

In August 1980, five years later, a sister of case 1 visited this hospital with the chief com-

^{*} 渡 捷一, 室積正人, 大作浩一: Brachydactyly との鑑別が困難であった Delta phalanx の姉妹例

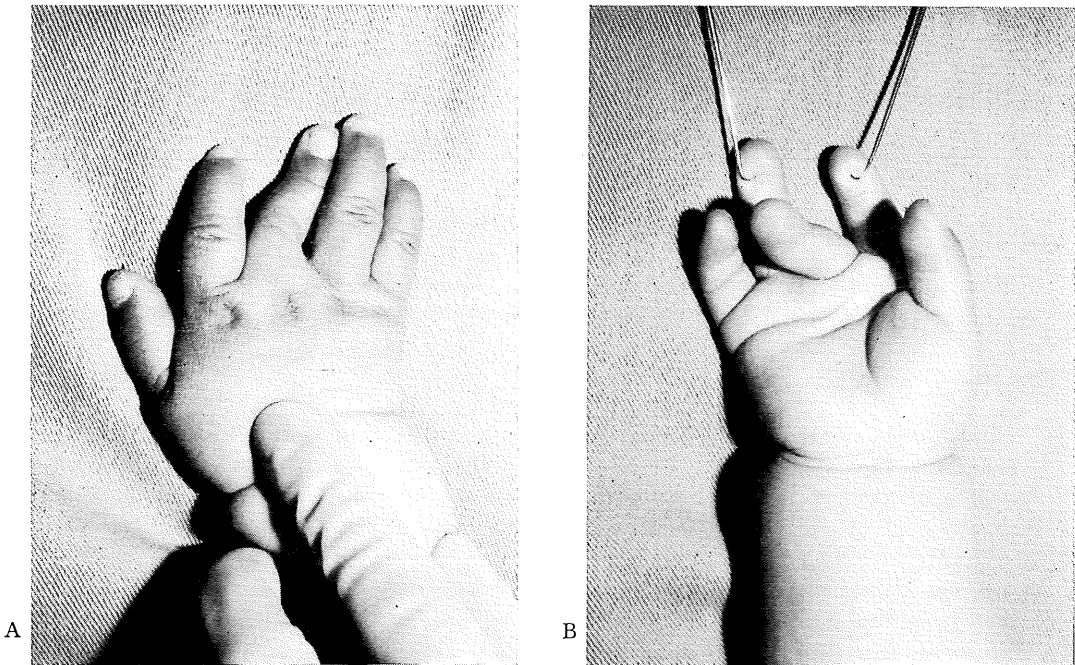


Fig. 1. External appearance of case 1 at admission. Because the two hands show mirror image, only the right hand is shown here.

A Dorsal view

The middle finger is short, ulnarly deviated in the vicinity of the MP joint and radially deviated at the DIP joint.

B Volar view

The middle finger shows marked overlapping with the ring finger in the position of flexion.



Fig. 2. Roentgenographic views of case 1 at admission. Both left and right hands show hyperphalangism, but the most proximal phalanx of the left hand in particular assumes the typical morphology of delta phalanx.



Fig. 3. Roentgenographic views of a case diagnosed as brachyhyperphalangism. Roentgenographic views of a case diagnosed as Vidal type brachyhyperphalangism are shown here as reference. They are very interesting compared with the views shown in Fig. 2.

plaint of deformity of fingers of both hands and it became a matter of controversy again. This case 2, the third child, was a girl 17 days old. No abnormality had been found in her 3-year-old older sister, the second child. As in case 1, no abnormality found in the history of pregnancy and delivery, and birth weight was 3110 g. Neither were pediatric abnormalities present.

The external appearance of case 2 is shown in Fig. 4. The left and right hands present a perfect mirror image. That is, the index finger and middle finger show brachydactyly, but the index finger is large and its nail is wide and large by far than nail of the thumb. And, both fingers present ulnar deviation from the vicinity of the MP joint. In the roentgenographic views, which are shown in Fig. 5, the development of numerous typical delta phalanges is apparent. The phalanx corresponding to the proximal phalanx and the middle phalanx of

both the left and right index fingers are delta phalanx, and two delta phalanges corresponding to the middle phalanx are seen. The index fingers present a bifid distal phalanx, so that digital ray fusion of two digital rays had obviously occurred. There is a little difference of middle fingers between the left and the right hand. That is, the right middle finger consists of four phalanges, is on the whole short, and presents brachyhyperphalangism, but the most proximal phalanx is definitely a delta phalanx morphologically. In the left hand, although a phalanx corresponding to the middle phalanx is present, epiphysis-like shadows are seen at both ends. And the phalanx corresponding to the proximal phalanx is, in this case also, a typical delta phalanx. (Fig. 4, Fig. 5)

As regards the diagnosis of case 2, the findings described above show that digital ray fusion is about to occur between the radial

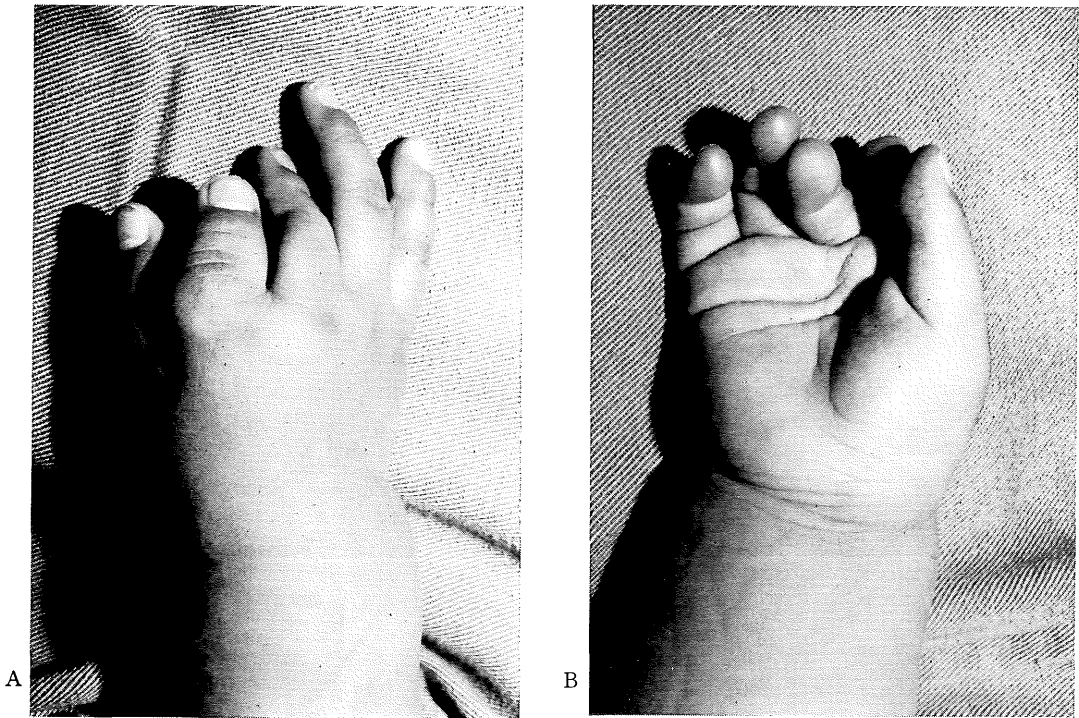


Fig. 4. External appearance of case 2 at admission. Views of the right hand are shown as the two hands present a mirror image.

A Dorsal View: Shortening and deformity of the index and middle finger are seen. The index finger is wide.

B Volar view: When the fingers are flexed, the ring finger overlaps the middle finger.



Fig. 5. Roentgenographic views of case 2. Both hands show trace of digital ray fusion in the index finger with appearance of three delta phalanges, and the distal phalanx is divided in two. In both middle fingers, the most proximal phalanx is a delta phalanx, and the right middle finger shows hypersegmentation.

polydactylous component of the middle finger and index finger. This was considered to be a characteristic findings of the cleft hand group. Since, in retrospect, the change in case 1 was also delta phalanx and in view of the fact that this was a case developing in a sister, it was considered appropriate to diagnose this as a deformity of cleft hand group.

These being sister cases, an investigation of the family tree was made. Although brachydactyly was suspected in a younger brother of the mother, this could not be confirmed and the genetic background remained unknown.

DISCUSSION

1. *Delta phalanx*

Delta phalanx is an abnormality of the phalanx which came to be well known after it was reported by Jones (1964)²⁾ and Watson and Boyes (1967)⁵⁾.

However, Jaeger and Refior (1971)¹⁾ and Wood and Flatt (1977)⁶⁾ raised objection to this designation, showing that the site of its develop-

ment is not limited to the phalanx but that it occurs also in the metacarpal and the metatarsal bone, and it is said that it is more universally referred to as triangular deformity of bone or delta bone at present. However, from the cases we experienced, this deformity is found at a preponderantly higher frequency in the phalanx and the frequency in the metacarpus and the metatarsus is so low that the development at these sites may rather be regarded as exceptional, so that the designation "delta phalanx" is not to be readily discarded.

Further, because this deformity presents a triangular shape morphologically, a broad interpretation is sometimes made that all phalanges presenting a triangular deformity are delta bones, but what is important is the morphology of the epiphyseal line which causes the triangular deformity. That is, an abnormality in which the epiphysis surrounds one side of the phalanx, namely, the two sides of the triangle, should be the basis for the diagnosis.

Various occasions have been reported for

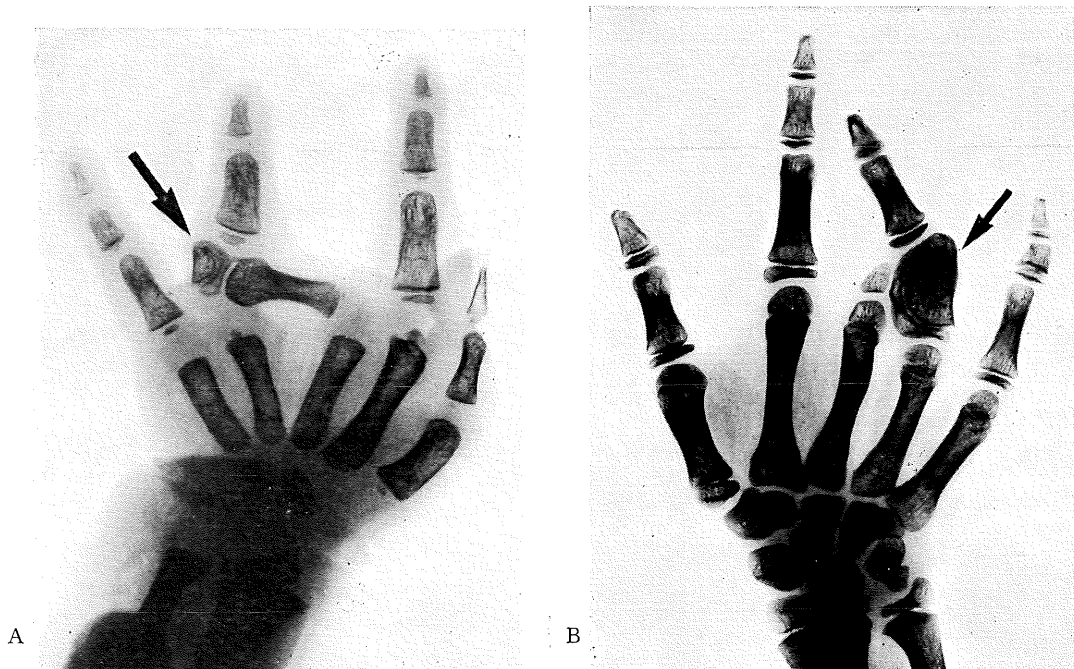


Fig. 6. Delta bones appearing in cleft hand group.

A On ray distal defect type. A cross bone is seen running ulnarly from the head of the third metacarpus. The proximal phalanx of the ring finger competing with this cross bone has become a delta bone.

B A similar case. The proximal phalanx of the ring finger cannot necessarily be said triangular, but the epiphyseal line shows a characteristic arrangement.

the development of this abnormality, ranging over numerous congenital abnormalities of the hand, such as polydactyly, syndactyly, cleft hand and ulnar ray deficiency. However, it is found on making a more careful observation and investigating when in the course of development of these abnormalities the present abnormality appears, that, as a common term, it appears in the process of digital ray fusion. Opinions agree that the deformity is frequently seen especially when digital ray fusion can be considered to have occurred in a part of polydactyly. Naturally, therefore, it is observed most frequently in the group of cleft hand, where the polydactylous change of the middle finger develop digital ray fusion with adjacent fingers⁴⁾. For example, the radial component of middle finger polydactyly develops digital ray fusion with the index finger and the ulnar component, with the ring finger, and at this time, one ray proximal defect of balanced type or imbalanced type develops depending on the size of the fused

polydactylous component. Delta bones formed in such a case are shown in Fig. 6 in A a cross bone runs ulnarly from the head of the third metacarpus and completes with the proximal phalanx of the ring finger, but the middle and distal phalanx and the index finger ray have already undergone smooth digital ray fusion and formed a single large digital ray. The delta bone shown with the arrow has a typical shape with two sides of the triangle surrounded by the epiphyseal line. A similar case is shown in Fig. 6-B. The phalanx indicated with the arrow is also surrounded by the epiphyseal line and is a typical delta bone, but it is not triangular as a whole. It appears that diagnosis of the present deformity should be based, not on the overall shape, but on the characteristic morphology of the epiphyseal line.

Delta bone found in the proximal phalanx has been described above, but this is frequently seen in the middle phalanx also, as shown in Fig. 7. Fig. 7-A shows delta bone seen in the

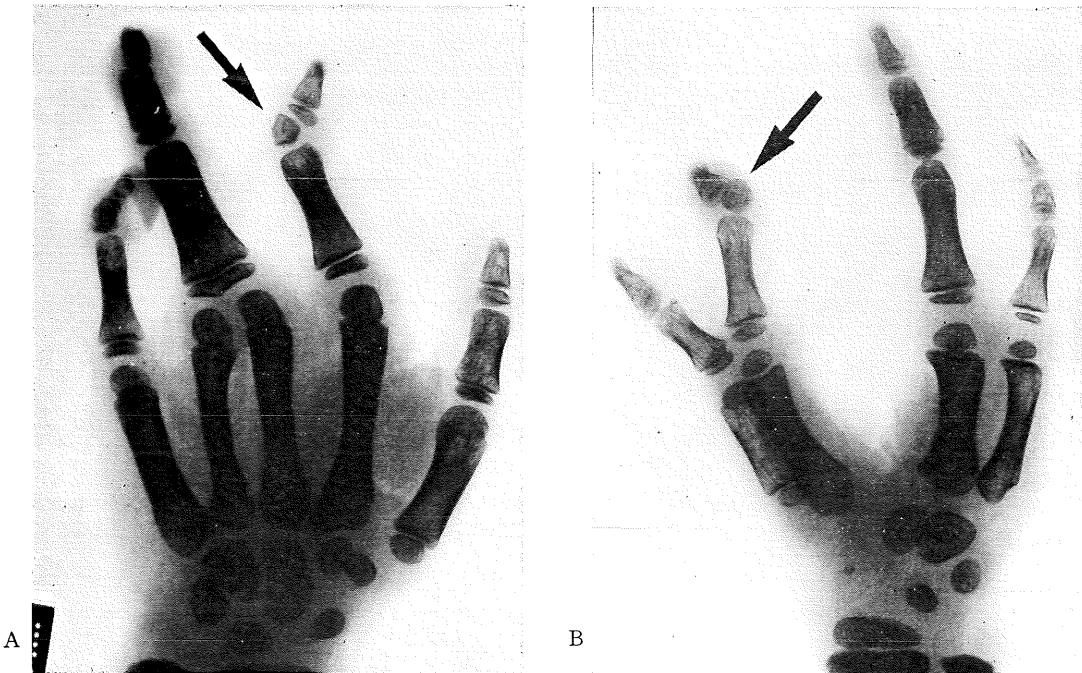


Fig. 7. Delta have appearing in cleft hand group

A Delta bone found in the middle phalanx of index finger. Presence of epiphyseal line is not clearly seen.

B Delta bone seen in the middle phalanx of index finger in one ray proximal defect type deformity

Though round, it can be identified as delta bone by the radial deviation of the DIP joint. As in A, presence of epiphyseal line is not clearly seen

middle phalanx of the index finger. In this case, the ring finger ray is large, and it is evident that digital ray fusion had taken place, but there being no hypertrophy of the bone in the digital phalanx of the index finger, the deformity possibly had occurred by digital ray fusion of only the digital rays of the middle finger and the ring finger. However, the transformation of the middle phalanx of the index finger into a delta bone, on the other hand, indicates that a digital ray fusion involving the index finger ray had also taken place. However, the delta bone of the middle phalanx does not show the characteristic deformity of the epiphyscal line. Similarly, a case with delta bone of middle phalanx of index finger is shown in

Fig. 7-B. The cleft hand is of the one ray proximal defect type. This delta bone cannot be said to be triangular. It is round, but it can be presumed to be a delta bone by the radial deviation of the DIP joint.

A deformity of the two ray defective type that is frequently found is triphalangeal transformation of the thumb which occurs due to digital ray fusion of the thumb ray and index finger ray. In this case, the phalanx corresponding to the middle phalanx generally because a delta bone as shown in Fig. 8. Quite the same phenomenon is to be observed when Wassel VII type thumb polydactyly undergoes fusion and becomes a triphalangeal thumb.

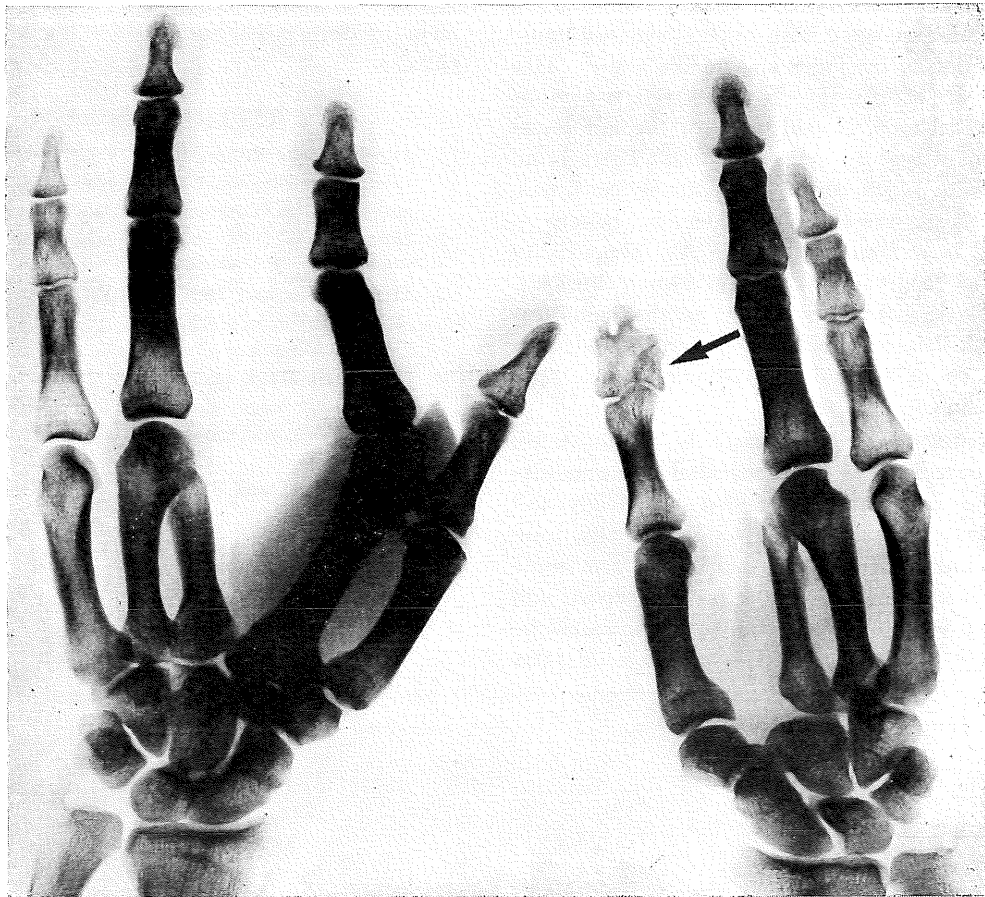


Fig. 8. Delta bone appearing in cleft hand group. A case of affection of both hands is shown. Cleft hand progresses by digital ray fusion of adjacent fingers, but in this case there is fusion of the thumb ray and index finger ray of the left hand and two ray defect is seen in the right hand. The delta bone indicated with the arrow shows a trace of this. The process by which Wassel VII type thumb polydactyly becomes a triphalangeal thumb is altogether the same.

2. *Brachydactyly*

An abnormality belonging to the category of brachydactyly whose pathology is the most profoundly studied in the field of hand surgery is symbrachydactyly which is complicated with syndactyly, but brachydactyly in the narrow sense has hardly ever been reported and such cases necessarily have to be sought for the most part in works in the field of genetics.

There are many types of brachydactyly in the narrow sense also, and the classification of Bell (1951)³⁾ is the classification that is frequently quoted in clinical genetics. By this classification, this abnormality is classified into five types from type A to type E, and type A is further subdivided into four types. Among these types, the authors' case 1 bears a close resemblance to type C, a close resemblance in that it is associated with hyperphalangial transformation of the index and middle finger. This type, an abnormality formerly referred to as the Vidal type, is said to be of the autosomal dominant type as to its heredity. There is no type with which our case 2 is directly consistent, but a case presenting an identical pattern is found in a family with type A-2 abnormality in the report of Temtamy and McKusick³⁾. Also, a report mentions that another name for type A-2 is deltaphalanx. These, viewed from the side of a hand surgeon, are not a little confusing.

As described above, there is a considerable difference in the way congenital abnormalities of the hand are understood genetically and the way they are understood surgically, and sometimes they are contradictory. The same is true of the cases of the present report, and it goes without saying that a review should be made in future whether there are some parts

to the concepts of brachydactyly and the cleft hand complex that we speak of which are as yet not established or whether their developmental mechanisms have come into being in common.

The cases described in the present report are cases developing in sisters and are naturally to be considered abnormalities of the same category, but establishment of diagnosis was difficult for brachydactyly and cleft hand complex because they both frequently show intrafamilial affection. However, the problems of the two abnormalities have come to our knowledge in the course of this review, and they have been reported.

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