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連體嬰兒面面觀

瓜堂堡 中山附設醫院 小兒外科主任

連體嬰的歷史背景

有關連體嬰的歷史,早在16世紀醫學文獻即有記載報告,而最早報告分割手術的是在西元1689年 König分割一對脐部連體嬰,他們僅是皮膚管相連而已。連體嬰經常會引起大家很大的興趣,不僅是因爲罕見,尤其是他們奇異的外觀及嘗試分割成獨立個體的挑戰,並提供了研究胚胎發生學的機轉及連體生活之生理情形一個不尋常的機會。現在連體嬰一出現,消息總是蓋不住,而且能否分割往往馬上成爲社會關心的話題,面對 // 兩個生命的未來 // ,醫學科技的大挑戰,同時他們會特別受到社會大衆的關懷和幫助,也正是人類對生命的尊嚴及兩個生命的重視。

目前文獻記載,全世界連體嬰出生后能存活一段日子的大約有四百對,其餘大部分是死胎或出生即死,而可以分割的在 1689~ 1962 間有 24 對分割,至1984爲止也僅有 85 對分割,綜觀國內,共有五對連體嬰分割,存活 8人,1974年省立台中醫院,首例成功分割第一對臍部連體嬰,後來僅存活一例,我們中山醫學院附設醫院共完全成功地參與了三對連體嬰的分割手術,包括最困難的忠仁、忠養坐骨連體嬰的分割,最近在本校附設醫院獨立完成一例臍部連體嬰的分割手術,這些都是極爲難得的成功經驗。

不願分割的連體嬰

至於連體嬰若不分割,是否可以活得一輩子呢?最有名的是西元1811年暹邏出生的一對連體嬰,後來 P.T. Barnum 將他們命名爲 "暹邏嬰(Siamese twin)",並帶到美國參加 "The Greatest Show on Earth"的演出,表演賺錢,更使他們富有一生,這對暹邏嬰 "Chang 及 Eng Bunker "的父親是中國人,母親有一半中國

血統,他們相連在一起不願被分割,過着很有趣的生活, 後來 Chang太太生了十個孩子, Eng太太也生了十二個小 孩,他們並且常常出去打獵,且是很出色的射擊手,同時 兩個人均活到六十三歲才一起死於肺炎。其實,我們實在 不希望連體嬰被養成表演賺錢的工具,因為那不合現代人 生活的意義。

成因及發生率

為何會生出連體嬰,我想除了上帝惡作劇外,尚無人能夠完全瞭解,全世界每年可能有六例以上可以分割的病例出現,但實際上很少,據統計:大約每五萬出生嬰兒有一對,70%是女性,黑人比白人發生率高,早期的文獻報告有60%是死胎,最近的統計報告,仍然有40%死胎,35%僅能活一天,其餘的25%出生后雖然能存活一段日子但能分割的却很少。連體嬰的發生,原因仍是未明,一般認為在受精卵着床,胚胎形第13~15天的不完全分裂所造成的不幸,而與母親的懷孕年齡無關,產前的超音波檢查,剖腹生產,可以減少母親因難產所造成的意外和不幸,同時,現代生殖科技的發達,優生保健更成為熱門話題。

應否分割與能否分割的問題

連體嬰的分割,除了複雜的醫學科技知識的認知外, 醫學倫理、宗教、法律理念的尊崇和配合均相當重要,尤 其對於分割后可能造成殘障,面臨存活的取捨,應否分割 的爭議等等,對於醫療本身可能是一種壓力和挑戰。首先 ,我們從醫學方面來探討影響成功分割的因素。由外科分 割手術的文獻報告中,我們知道,分割手術成功的因素有 連結部位的解剖複雜性,共有器官的程度及形式,倂發畸





。。連若後分爭,順科硬外順當男連兩手性者複理部,

新生

使孩婴討的:治療

圖一;a,b 附設最近分割成功的一例臍部連體嬰。由b.的 X-Ray 顯示,並無共有生命器官。

昌一

形及處理方式。(一臍部連體嬰:分割的病例最多,成功例 也最多, 因為連結情形最少,常常無共有生命器官, 有的 僅在肝臟之間有纖維帶存在而已我們醫院最近分割的一例 情況相似,然而主要的分割問題在於肝臟有否嚴重相連。 口胸部連體嬰:次多的一種,但分割成功的並不多,到目 前爲止,心臟相連,使得分割成爲不可能。縱使有分開的 心臟,因缺少胸骨,使得在術后因胸壁的不穩定而引起無 法克服的呼吸問題。 < < > 坐骨連體嬰: 並不多,可能是可分 割的連體嬰中最複雜的,文獻上只有42病例報告,但只有 六例分割成功,其中1966年在美國塩湖城Eades的首例成 功,1970年忠仁、忠義連體兄弟是全世界首例三隻脚坐骨 連體男嬰分割手術成功,兩者均存活,由於分割成功只有 一半機會,當時,醫師的壓力很大,經過兩年半的公開討 論,手術時間超過12小時,得以完全成功,參加手術的醫 護人員立獲 蔣經國總統召見加勉,在台灣醫界帶來無限 鼓勵及最高的榮譽。四臀部連體嬰:危險性小,除非共有 一個共同 萬椎管, 此種連體 嬰除非犧牲讓其中一個或二個 ,世界已有分割成功病例,兩個完整的個體在顱骨穹隆成 鏡像部位相連,可能有或無腦及腦膜相連,國內迄今尚無 此病例存活出現。

分割的抉擇與法律問題

連體嬰相連的複雜性如何?最近由於 CT, MR I, 超晉 波及 X 光檢查的普遍應用,幾乎在極短的一天內就可清楚 的了解各相連器官的關係位置及情形,提供能否分割及分割難度的寶貴資料。1986年我們中山接到了另一對四隻脚坐骨連體嬰,其中一位正常,另一位有腦小,心臟有先天性缺損,心衰竭,及血液幾乎靠正常側輸送,由檢查判斷知道無法存活兩位,緊急處理可能要犧牲缺陷的一位來救 活正常的一位,但表面上這又會牽涉到法律問題,最後,在檢查完竣轉送台大醫院分割救活其中正常的一位,這也算是完全成功分割的病例。

分割的標準

分割手術是爲了連體嬰兒能各自獨立生活的唯一途徑 ,1968年 Prof. Gans 等主張四種分割標準: → 當連體嬰 中的某一個生下卽死亡或其生命情況可危及另一個生命時 。口當有先天性畸型而無法維持生命且須立卽矯正治療時。曰當連體嬰已接受完整的檢查且適宜分割手術時。四當連體嬰的相連部分發生了重大的病變時須立卽分割手術。四當若相連部位簡單,較無生命危險的器官,組織,而且分割後,兩者均能擁有各自獨立完整的個體及正常人的生活,分割手術僅是醫學技術層面的討論和準備,較爲單純及無爭議性。但是,對於相連部位複雜,合倂畸型,應否分割,兩者能否同時存活,術后有否殘障而需社會長期輔助照願等複雜問題,在術前宜從醫學、宗教、法律、新生兒外科倫理等方面作評估,執行分割手術的醫院,除了必要的硬體設備外,各專門科系人才的能力要求和配合,尤其是外科,麻醉科,小兒科,護理工作人員的團除合作,才是順利達成分割手術的重要因素。

新生兒外科倫理的探討

每個人都希望自己生下的小孩健康快樂,醫學的進步使以前無法治療的變爲可能,使許多先天性缺陷或畸型的孩子,獲得了幾乎完全與正常人的相同外貌和功能,連體嬰要不要救治,應否分割,也牽涉到新生兒外科倫理的探討和醫學倫理的判斷。根據,Rickham 對於先天性畸型的分類判定標準分爲五群,可以提供吾人參考:(一)第一群:可以用外科治療,並能完全治癒者。(二)第二群:用外科治療後,可能殘留少許障礙,但可以在社會上正常的生活

者。曰第三群:雖然接受外科治療,但因爲嚴重的障礙,致無法與正常人一樣在社會上生活者,就如坐骨連體嬰忠仁、忠義只有三隻脚,分割後僅能各自擁有一隻正常的脚,成爲先天性殘障者,此時要看整個社會是否有愛心能接納,我們的社會大衆爲忠仁、忠義出錢,同時分割后,從身體、心理復健至敎育的啓蒙,均有善心和愛心的慈善機構來幫忙收容照顧,目前忠仁忠義已十三歲,就讀國中,智能均正常,成績也很好。四第四群:屬於第一至第三群,智商低,但經敎育,可稍許改善者。因第五群:屬於第一至第三群,但智商極低,幾近於所謂植物人者,救治的希望少,也無實際的生活意義。

醫療、法律和社會責任觀

站在法律的觀點,醫師只要盡自己的所能幫助病患改善痛苦,連體畸型兒和他們的父母都是受害者,社會應共同分擔他們的痛苦。

我們 "中山" 很幸運地在最近十三年來陸續接到三例 連體嬰兒,相連情況雖然不同,由於醫院的成長和進步, 三對病例,均在我們醫院妥善的安排檢查,照顧及參與下 ,兩對後來轉送台大醫院分割成功。同時,去年九月廿六 日在本校附設醫院獨立完成的臍部連體嬰在新生兒期分割 手術,更可肯定我們醫院各方面的努力和成就,也是值得 慶賀的歷史一頁。

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SOLEUS MUSCLE FLAP FOR COVERAG OF LEG DEFECT

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SUMMARY

We resurfaced defects of the middle one third of the leg with a soleus muscle flap in eight patients and have obtained favorable results for coverage of these open wounds. There was one complication wherein there was a 20 degree extension lag of the knee and a 15 degree dorsiflexion loss fo the foot, This muscle is expendable and very versatile in selected cases. We describe its use as a split muscle transfer to better suit the wound size or contour.

A variety of surgical methods have been used for coverage of soft tissue defects of the legs. These includ skin grafting, local cutaneous flaps (Recalde Rocha, J.F. , et al., 1987), muscle flaps (Dibbel, D. G., et al., 1980; Kojima, T., 1980), fasciocutaneous flaps (Cormack, G.D., et al., 1984) and free flaps (Serafin. D., et al., 1983). If patients have exposed bone, skin grafting is ineffective and a flap is needed. Local flaps should be considered first since distant tissue transfers are more time-consuming, (Morris and Buchan, 1978; Serafin, et al., 1977). In our experience , twenty-two local muscle flaps were used in seventeen patients. This included eight soleus muscles with favorable results. We

feel that the soleus muscle is still an important flap for coverage of defects of the middle one third of the leg.

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OPERATIVE METHOD

With the patients in the supine position, the exposed, necrotic tissue in the defect is debrided. The dissection is done using a medial longitudinal incision a little anterior to the Achilles tendon. After the fascia is incised longitudinally, the planes superficial and deep to the soleus muscle are identified. Combining sharp and blunt dissection, the dissection is continued up to the major pedicles and down to the malleolar level. If the defect is long, the muscle can be carefully longitudinally split in the midline to meet this need (Fig. 1). Usually we ligate one or two pedicles, arising from peroneal artery to facilitate the transposition. After cutting the tendon at the malleolar level, the muscle is transposed into the defect. A split-thickness skin graft is applied to the transposed muscle.

PATIENTS

The soleus muscle was used for eight patients (six males and two females, ranging from 19 years to 55 years of age). All of the cases had sustained traffic trauma within six months except for one case of

70

an unstable wound following a snake bite 37 years earlier, The soleus muscle alone was used in six cases. The soleus and peroneus brevis was used in one case and the soleus and extensor digitorum longus was used in one case (Table 1).

CASE EXAMPLE(1)

19 year old male seen in March, 1985, presenting history of a traffic accident four months previously, initially treated on a local orthopaedic service. He experienced malignant hyperthermia intraoperatively during a prior operation. He was also treated with traditional Chinese medicine. On physical examination, he was illlooking, had lost weight and was mal-nourished. The exposed bone of the tibia was 18 cm long (Fig. 1A). We used a soleus muscle flap with longitudinal splitting to cover the defect via a medial approach (Fig. 1B). The postoperative course was uneventful (Fig. 1C). He wore a long leg cast for three months and a patella tendon bearing cast for the following six months. He returned to work as a car driver 12 months after flap surgery (Fig. 1D).

CASE EXAMPLE-(2)

(2) A 50 year old house-wife was bitten by a snake 37 years previously. This resulted in unstable scar with bony erosion over the right anterior tibial area (Fig. 2A). When first seen she was hospitalized in a local private hospital without microsurgical capabllity. At this hospital, the wound was covered with a soleus muscle (transposed via a medial approach) and skin graft after being debrided. Coverage was achieved in ten days. The defect remained well healed at one and a half years follow-up (Fig. 2B).

RESULTS

In this series our postoperative followup period was from 12 months to 42 months (average 24.5 months). The result was satisfactory in seven cases. One case had a residual knee extension loss of 20 degrees and a foot dorsiflexion loss of 15 degrees. Take of the skin graft on the muscle flap was complete in ten days. The wounds were

resurfaced with durable tissue.

DISCUSSION

Recent reports indicate favorable results for resurfacing leg defects by debridment of the wound, stabilization of the fractures and appropriate coverage of the fracture site (asko-Seljavaaras, et al., 1985). Microsurgical free flaps are now more commonly used for this resurfacing (Serafin, D., et al., 1980;1983). We also use free flaps to a great degree. The use of conventional muscle flaps to achieve favorable results for leg resurfacing have been

previously reported for open fracture of the tibia (Kojima, T., 1980), chronic osteomyelitis (Ger, et al., 1977; Mathes, S.J., et al., 1982), bone grafting and tumor ablation (Kojima, T., 1980). The use of a muscle flap has many merits:(1) The operating time is short, sometimes less than one hour. (2) The operation can be finished without microsurgical techniques. (3) The muscle flap surgery can be done concomitantly with bone grafting and it is of great value when there is bone infection (Kojima, T., 1984; Mathes, S.T., 1984).

In 1966 Ger introduced the soleus muscle flap for open tibial fractures. Subsequently other surgeons have reported their experience using this muscle. The muscle has the following features (1) The flap has robust blood supply from the popliteal artery, posterior tibial artery and peroneal artery. (2) It is easy to be transposed from either the medial or lateral approach (Mathes, S.J. and Nahai, F., 1982). (3) The muscle volume is large and it is long so can be split for different sized

TABLE I

TABLE 1 Muscle for reconstruction				
Soleus	6			
Soleus and				
Extensor digitorum longus Soleus and Peroneus brevis	1			
Total	8			

defect. (4) If the defect is on the medial side of the middle third of the leg, a distally based soleus muscle can be of use (Fayman, M.S., et al., 1987; Mathes, et al., 1980).

Of the 8 patients we report, none have developed bony exposure. In our 1 case with a complication, we think the leg trauma was more severe and treated too late. This patient had extensive rehabilitation but still shows a functional loss

The other patients are currently ambulatory with no wounds. We feel that better resulas are likely if the patients are treated as soon as possible after trauma.

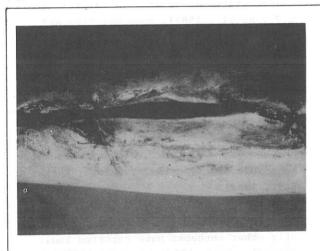
REFERENCE.

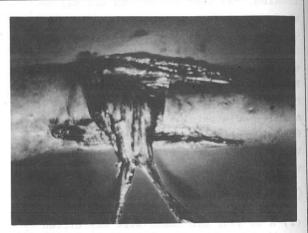
Asko-Seljavaaras, S., Slatis, P., Kannisto, M., and Sundell, B.(1985).
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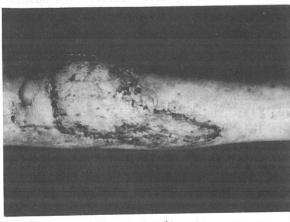
TABL

Cormack, G.C., and Lamberty, B.G. (
 1984). Fasciocutaneous vessels: their

FIG I







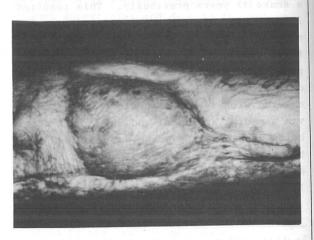


Figure 1(Right, foot to right)

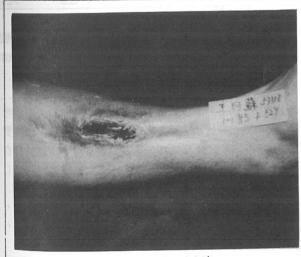
A.Left Above. When admitted, tibial exposure 18cm.

- B.Right Above. Soleus muscle flap with longitudinal splitting.
- C.Left Below.Postoperatively,ten days.
- D.Right Below.Postoperatively,12 month follow-up.wound healed completely.

TABLE 2

Case	Age Sex	Caab	ollow up eriod	Wound condition	Final result
	40m		42m	soft tissue defect 8×10 cm, tibial exposure 2×5 cm	helling
2 2200	35m	motorcycle	40m	compound tibial and fibula fracture, exposed tibia $8\mathrm{cm}$	healing
_	18f		32m	compound tibial and fibula fracture, exposed tibia $6\mathrm{cm}$	healing
	30m		20m	bilateral tibial and fibula fracture, exposed tibia $4\mathrm{cm}$	healing
5	30m		18m	compound tibia and fibula fracture, exposed tibia 7cm	healing
	55f	snake bite	18m	osteitis, unstable scar with hischarge	healing
7	19m	motorcycle	12m	compound tibia and fibula fracture, exposed tibia 18cm	knee extension lag 20°,Ankle
	6449 100				dorsiflextion lag 15°
8	40m	motorcycle	12m	compound tibia and fibula fracture, tibia exposure 6cm	healing

FIG 2



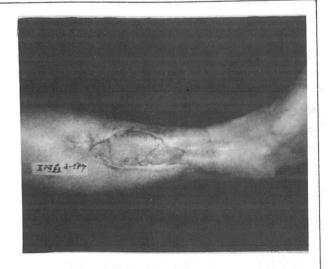


Figure 2 (Right leg; foot to right)

A.Unstable scar with chronic discharge for 37 years.

B.Postoperative follow-up $1\frac{1}{2}$ years.

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