Tongue-Lip Adhesion in the Management of Pierre Robin Sequence with Airway Obstruction: Technique and Outcome

Faye Huang, MD; Lun-Jou Lo[1], MD; Yu-Ray Chen[1], MD; Johnson C. Yang, MD; Chen -Kuang Niu[2]; MD, Mei-Yung Chung[2], MD

Background: Airway obstruction and feeding difficulty can occur in patients born with Pierre Robin sequence. In select patients with pronounced micrognathia, surgical intervention to relieve the airway obstruction is necessary. The surgical indications and appropriate surgical procedure continue to have a great deal of controversy. The purpose of this study was to evaluate our experience of tongue-lip adhesion in the management of upper airway obstruction associated with Pierre Robin sequence.

Methods: From March 1995 through May 2002, a total of 14 patients with Pierre Robin sequence, who were admitted to the pediatric neonatal intensive care unit either with prolong intubation, poor body weight gain, or repeated airway infection, underwent tongue-lip adhesion (TLA). The operation was performed by raising mucosa flaps and approximated the muscles between tongue and lower lip. Retention sutures were used. The patients were evaluated for clinical responses.

Results: Our successful rate with tongue-lip adhesion was 70%. Ten of the 14 patients showed clinical improvements including extubation of the endotraccheal tube, body weight gain, return for home care, reduced episodes of respiratory infection, and improvement in O2 saturation and blood gas. The surgical procedure was simple to perform without major complications.

Conclusions: This retrospective review showed that with a thorough preoperative airway evaluation, TLA could be successfully used to treat select patients with Pierre Robin sequence suffering from severe upper airway obstruction. Thus, TLA should be first considered when surgical relief of airway obstruction is indicated and when obstruction is limited to the classic tongue base obstruction type.

(Chang Gung Med J 2005;28:90-6)

Key words: tongue-lip adhesion, Pierre Robin sequence, airway obstruction.

The triad of glossoptosis, micrognathia, and cleft palate is known as Pierre Robin Sequence (PRS).[1] The concept of "sequence" suggests that one anomaly causes subsequent anomalies, and micrognathia is believed to be the inciting anomaly in patients with PRS.[1,2] Airway obstruction and

From the Department of Plastic and Reconstructive Surgery, Chang Gung Memorial Hospital, Kaohsiung; [1]Craniofacial Center, Department of Plastic and Reconstructive Surgery, Chang Gung Memorial Hospital, Taipei; [2]Department of Pediatrics, Chang Gung Memorial Hospital, Kaohsiung.

Received: Oct. 27, 2004; Accepted: Dec. 21, 2004

Address for reprints: Dr. Faye Huang, Department of Plastic Surgery, Chang Gung Memorial Hospital. No. 123, Dabi Rd., Niausung Shiang, Kaohsiung, Taiwan 833, R.O.C. Tel: 886-7-7317123 ext. 8002; Fax: 886-7-7317123 ext. 8004; E-mail: spencer-faye@cgmh.org.tw

feeding difficulties arise and the severity of the problems vary. PRS can be life threatening during the neonatal period with the onset of airway obstruction, which can occur at any time right after birth. If left untreated, prolonged airway obstruction can lead to acute or chronic hypoxia, cyanosis, apnea episodes, aspiration, respiratory tract infection, feeding difficulties, malnutrition, and failure to thrive. Subsequent complications of chronic hypoxia are chronic carbon dioxide retention, elevated pulmonary vascular resistance, cor pulmonale, right heart failure, and cerebral hypoxia.[1,3,4] It is therefore important to identity and treat airway obstruction as soon as possible. The treatment protocol for PRS usually starts with conservative and positional management. Pharyngeal airway appliance or tracheal intubation is used to keep airway patent. Surgical intervention is considered for a patient with prolonged use of an appliance or intubation, failure of conservative management, and repeated problems related to airway obstruction. Surgical methods include tracheostomy, tongue-lip adhesion (TLA), mandibular distraction, and others. However, the selection of surgical methods differs among surgeons and patient care centers.

TLA is a simple surgical procedure in which the surgeon sutures the tongue anteriorly to the lower lip and opens up oropharyngeal airway space as the tongue base is pulled forward. Techniques of TLA were introduced in the 1940s and have been modified.[5-8] The application of TLA was controversial, mainly because of the concern regarding its effectiveness.[9-11] As the tracheostomy during the infant stage may include the need for complicated care and possibility of morbidity, TLA has been employed as an initial step when surgical intervention for airway management is indicated at the Chang Gung Craniofacial Center.[12, 13] Secure adhesion procedures were used to prevent dehiscence of TLA. In this retrospective study, we report our experiences of TLA on 14 consecutive PRS patients with airway obstruction that did not respond to conservative management.

METHODS

From June 1995 through May 2002, a total of 14 patients were identified with PRS and were treated with tongue-to-lip procedures. The characteristic appearances of small and recessed chins were present. Thirteen full-term newborns with one premature infant were enrolled in this study. The mean body weight was 2894.9 gram with the range between 1960 g to 4100 g. The mean age at the time TLA was performed was 87.07 days old with the range between 18 days to 348 days. Two patients were initially treated with tracheotomy at other institutions. Twelve of the patients had isolated cleft palates and two patients had normal palates. Other associated anomalies included congenital heart disease, brain atrophy, and Hirschsprung's disease. Twelve of the 14 patients were admitted to the pediatric intensive care unit, with prolonged use of oropharyngeal airway appliance or tracheal intubation, difficult feeding, poor body weight gain, and repeated respiratory infections. Our initial treatment for the patients with PRS was conservative treatment which including position, instrumental airway maintenance, oxygen supply, nutritional support, antibiotics, and chest care. When conservative treatment failed to treat the acute airway obstruction, surgical treatment was performed. Surgical intervention was judged necessary by both pediatricians and surgeons from the presenting conditions, medical history, and results of nasopharyngoscopy. When there was no other associated airway problems or neurological diseases, TLA was the first step airway management proposed and consented to by the parents. In two patients, tracheostomy had been performed in other hospitals, and TLA procedures were accepted in order to facilitate decanulation and home care.

The TLA operation was performed under general anesthesia. Orotracheal intubation for anesthesia was applied in the 12 patients without a prior tracheostomy, and intubation through the tracheostomy was used in the remaining two patients. Traction sutures at the lateral aspect of the tongue were performed on both sides for pulling out the tongue. The tongue frenulum was released by direct electric coagulation if it restricted the outward movement of the tongue. An unrestrained tongue protrusion from the root of the tongue was ensured. The ventral surface of the tongue was pulled anteriorly to contact the lower lip mucosa. A slight tension was exercised with this action in order to move the tongue base forward to release the pharyngeal airway. A contact area was estimated for muscle adhesion. An inferior-base mucosa flap about 20 x 8 mm from the lower lip and

a superior-base mucosa flap at same size from the ventral surface of tongue were elevated, exposing the underlying muscle of the lower lip and tongue. The lower lip mucosa flap was flipped backward across the gum and sutured to the lower edge of the tongue wound. Solid muscle-to-muscle approximation between the tongue and lower lip was achieved using three to four 4-0 PDS sutures. The sutures were tied at the same time after all were placed. Then the tongue mucosa flap was flipped forward and sutured to the superior edge of the lower lip wound, covering the tongue-lip muscle sutures (Fig. 1). Finally two retention sutures using 3-0 Nylon were anchored near the tongue base, brought out through chin skin surface, and tied over silicone buttons (Fig. 2). The postoperative care was non-specific.

The retension sutures were kept for 2 weeks and used to protect the muscle adhesion for smooth healing. The patients were evaluated for clinical responses, including feeding, oxygen saturation, and body weight gain. If improvement was observed during the follow-up examinations, a trial of removing endotracheal tube was performed.

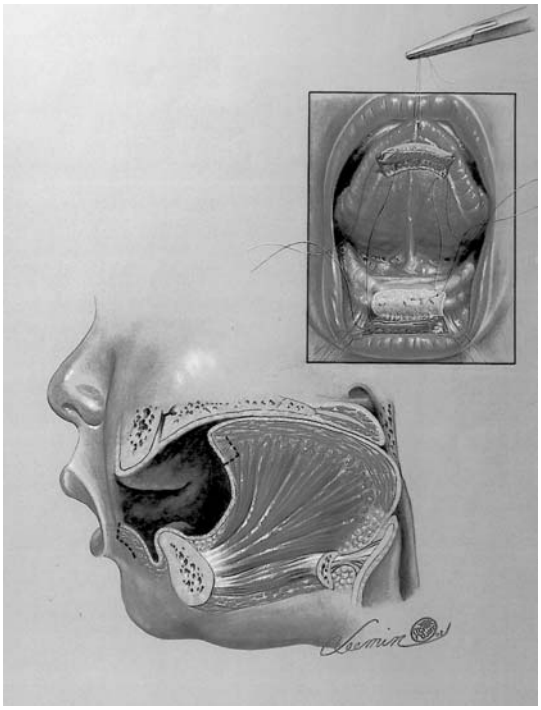

Fig. 1 The lower lip mucosa flap was flipped backward across the gum and sutured to the lower edge of the tongue wound. Solid muscle-to-muscle approximation between tongue and lower lip was achieved using three to four 4-0 PDS sutures. The sutures were tied the same time after all were placed. Then the tongue mucosa flap was flipped forward and sutured to the superior edge of lower lip wound, covering the tongue-lip muscle sutures.

RESULTS

Fourteen patients were identified with PRS and were treated with TLA. Patients remained on intubation after the operation for a period of 5 to 12 days. Retention sutures were kept in place for 12-14 days. Four patients had wound dehiscence, and all four had received a second procedure of the tongue-lip adhesion. Our success rate, which also included patients who needed a second procedure of TLA, was 71% (10/14), in terms of successful airway management. Patients with successful airway management showed clinical improvements such as body weight gain, return to home care, marked reduction in the episodes of respiratory infection, and improvement in both arterial blood gas and O2 saturation. In our series, five patients needed further surgical treatments including a second procedure of the TLA and three needed tracheostomies. One of the two patients

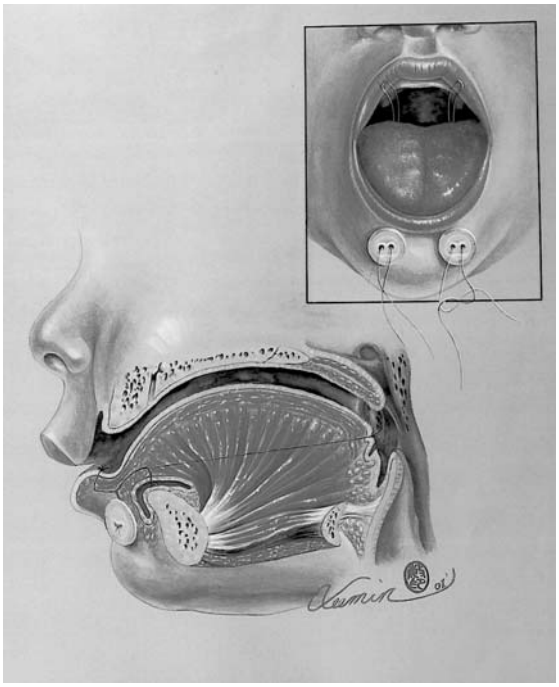

Fig. 2 Two retention sutures using 3-0 Nylon were anchored near the tongue base, brought out through chin skin surface, and tied over silicone buttons.

who had tracheotomy done elsewhere was successfully decannulated after TLA procedure. The other patient with the prior tracheostomy remained on tracheostomy despite of the performance of TLA.

After the TLA procedure, 3 patients had prolonged nasograstric tube feeding. Feeding difficulties may persist from 1 to 3 months. Most of our patients went to bottle-feeding without difficulties. Seven of the 14 patients had TLA performed by the junior author, and the timing for the division for TLA and palate repair was at 12 months old. Five of the patients underwent the division of tongue-lip adhesion and had palate repair at 12 months of age. Two patients without cleft palates had division of TLA at 12 months of age. No patient demonstrated obstructive apnea after palate repair and concomitant take down of the tongue-lip adhesion at 12 months old. The remaining 7 patients had palate repair at 12 months old or older and take down of TLA at 18 months or 3 to 6 months after palate repair. In the group of patients who had division and palate repair performed at different times, TLA was performed by the senior author. Division of the TLA and palate repair was performed either at the same time or at two different times, as long as the patient showed a normal weight gain, without repeated airway obstruction or infection and without feeding difficulties before the palate repair or the division of the TLA. There were no no death in our patients. The mean hospital staying was 58 days with the range of 27 days to 194 days.

Three PRS patients required further tracheotomy. One patient who had a prior tracheotomy and remained on tracheotomy, had laryngomalacia and tracheomalacia, which was confirmed by nasopharyngoscopy. Another patient who needed tracheotomy was associated with multiple brain infarctions. The patient who needed tracheostomy and the one who remained on tracheostomy did not receive nasopharyngoscopy evaluation prior to TLA because it was not a routine procedure at the time of their admissions.

DISCUSSION

Most of the infants with Pierre Robin sequence are successfully managed using non-surgical techniques. However in neonates with pronounced micrognathia, failure to thrive due to chronic airway obstruction, or severe respiratory distress, surgical intervention to relieve the obstruction is necessary. This may take the form of tongue-lip adhesion, distraction osteogenesis, or tracheotomy. Due to the great variation and severity of airway obstruction and feeding difficulties encountered in these patients, there is no consensus with regard to the choice of method for surgical airway management.[14] Using surgical approaches that are less invasive with fewer long term complications than tracheotomy is our preferred treatment of choice. The results of this retrospective study show that TLA effectively released airway obstruction in 71% of patients. Thus, tracheostomy can be avoided in the majority of patients who required surgical airway management.

Glossoptosis is not the only contributor to upper airway obstruction in children presenting with PRS.[15-18] Factors that cause upper airway obstruction in infants with PRS are often the primary features of the associated syndrome and could include skull base anomalies, pharyngeal hypotonia, nasal airway constriction, lower airway abnormalities, and central nervous system disorders.[15,16] For this reason, timely diagnosis and accurate morphologic and endoscopic work-ups are of extreme importance. The treatment options essentially depend on the location, severity, and mechanism of airway obstruction. Depending on individual status, the appropriate methods of management of PRS patients during the neonatal period include prone positioning, pediatric nasopharyngeal airway, endotracheal intubation, mandibular traction, orthopedic appliance, tongue-to-lip adhesion, tracheotomy, and distraction osteogenesis.[4,19,20] Conservative management should always be performed prior to surgical treatment. Weight gain, arterial blood gas, pulse oximeter, apnea monitor, and respiratory rates of the patients were monitored as references to determine the effects of the specific treatment. Nasopharyngeal airway has been useful in relieving the airway obstruction of patients with PRS. Although using nasopharyngeal airway seems to be a simple technique, the placement of nasopharyngeal tube at an appropriate level is a challenge. The nasopharyngeal tube is an unstable mode of therapy especially when it needs to be employed for a long period of time.[9] Prolonged use of the nasopharyngeal airway or endotracheal intubation constitutes the need of surgical airway management.

Tracheotomy should be a primary consideration

in patients with multiple areas of airway obstruction, significant secondary respiratory abnormalities, and central apnea.[14] Tracheotomy was avoided by most surgeons mostly due to longer hospital stay, parental consent, and the demand for home care. Tracheostomy has greater morbidity, especially in newborns.[12,13] Complications such as tracheal stenosis, granuloma formation, and esophago-tracheal fistula were other reasons for using tracheotomy as the last resort for the management of airway in PRS. Although mandibular distraction osteogenesis may provide a definitive structural resolution of micrognathia, complications with distraction osteogenesis have included damage to the teeth buds, injury to the inferior alveolar nerve, and unsightly facial scars.[2,21] Distraction osteogenesis is technically more difficult than other alternatives and requires good compliance from the parents.[2]

The concept of tongue-lip adhesion for relief of obstructive apnea associated with PRS was first popularized by Douglas in 1946.[5] Many modifications of the procedures of tongue-lip adhesion have been published including Routlege in 1960, Randall in 1977, and more recently Argamaso in 1992.[6] In our series, three of the four dehiscence from tongue-lip adhesions occurred during the early years when the technique was being developed. The incidence of complications and the failure rate of tongue-lip adhesions has decreased as the technical modifications have improved and with increased experience in performing the procedure.[6,22] The concept of strong muscle-to-muscle sutures arose from our experience of performing adhesion cheiloplasty for patients with wide cleft lip and palate.

Based on the study results by Sher et al, by using flexible fiberoptic nasopharyngoscopy in a review of 53 infants with PRS, four types of airway obstructions were found.[23] In type I, the obstruction is caused by posterior movement of the tongue against the posterior pharyngeal wall and was the most common. In the cases of Type II, the obstruction is due to the posterior and the superior displacement of the tongue promoting contact between the tongue, velum, and pharyngeal wall in the superior oropharynx. Type III obstruction is a pharyngeal obstruction, caused by prolapse of the medial wall of the pharynx. Type IV obstruction is due to the constriction of the pharynx in a circular manner by movement of the tongue and both lateral pharyngeal wall. Only for cases of type I obstruction, without any other associated anomalies, is TLA the treatment of choice.[6,10] In addition, it must be determined that the upper airway obstruction can be fully resolved by positioning the tongue forward. As for Type II, III, and IV, they do not respond well to TLA because other factors contributing to apnea and tracheostomy may be indicated.

Although TLA is a simple procedure, complications can still occur. Reported complications involving all forms of TLA are button and suture cutting through the tongue, wound dehiscence, injury to Wharton's duct, scar formation on lip, chin, and floor of mouth, feeding problems, epiglottis tethering leading aspiration, and dental abnormalities.[10,11,20] Using our TLA method, we have found that the number of wound dehiscence decreased, and there were no unsightly scarring, feeding problems, or dental abnormalities.

Another important cause of failure of TLA is poor patient selection, as in the presence of laryngomalasia and tracheomalasia that occurred in this series. Careful assessment including nasopharyngoscopy provides helpful information for the proper selection of surgical modality and preventing treatment failure. Nasopharyngoscopy is now a routine in our center and should be performed to rule out other upper or lower airway problems and to see the obstruction type prior to the performance of TLA.

In conclusion, airway obstruction in PRS involves multiple factors, there is no single treatment that will resolve all cases.[6,17,23] Appropriate airway management and feeding programs must be tailor-made for each PRS patient. A thorough preoperative airway evaluation is therefore of paramount importance before any treatment is implemented to rule out any underlying airway pathology and concurrent neurological diseases. As demonstrated in our review, tongue-to-lip adhesion is a simple surgical procedure without major or long-term complications. In the classic base-of-tongue obstruction, i.e., Sher's type I obstruction without other associated airway abnormalities, TLA should be considered first when surgical relief is indicated.[6,15]

REFERENCES

1. Tomaski SM, Howard GH, Aaal HM. Airway obstruction in the Pierre Robin Sequence. Laryngoscope

1995;105:111-4.
2. St-Hilaire H, Buchbinder D. Maxillofacial pathology and management of Pierre Robin Sequence. Otolaryng Clin N Am 2000;33:1241-56.
3. Dykes EH, Raine PAM, Arthur DS, Drainer IK, Young DG. Pierre Robin Syndrome and pulmonary Hypertension, J Pediatr Surg 1985;20:49-52.
4. Benjamin B, Walker P. Management of airway obstruction in the Pierre Robin sequence. Int J Pediatr Otorhi 1991;22:29-37.
5. Douglas, B. The treatment of micrognathia associated with obstruction by a plastic procedure. Plast Reconstr Surg 1946;1:300-8.
6. Argamaso RV. Glossopexy for upper airway obstruction in Robin sequence. Cleft Palate Cran J 1992;29:232-8.
7. Ryan RF, Longenecker CG, Krust L, Vincent RW. Anterior fixation of the tongue, a modification of Douglas and Routledge techniques. Plast Reconstr Surg 1963;32:318-21.
8. Parsons RW, Smith DJ. A modified tongue-lip adhesion for Robin anomaly. Cleft Palate J 1980;17:144-7.
9. Augarten A, Sagy M, Yahav J, Barzilay Z. Management of upper airway obstruction in the Pierre Robin Syndrome. Br J Oral Max Surg 1990;28:105-8.
10. Bath AP, Bull PD. Management of upper airway obstruction in Pierre Robin sequence. J Laryngol Otol 1997;111:1155-7.
11. Caouette-Laberge L, Plamondon C, Larocque Y. Subperiosteal release of the floor of the mouth in Pierre Robin sequence: experience with 12 cases. Cleft Palate Cran J 1996;33:468-72.
12. Gianoli GJ, Miller RH, Guarisco JL. Tracheotomy in the first year of life. Ann Oto Rhinol Laryn 1990;99:896-901.
13. Weissler MC. Tracheostomy and intubation. In: Bailey BJ, Calhoun KH, eds. Head and Neck Surgery-Otolaryngology. 3rd ed. Philadelphia: Lippincott Williams & Wilkins Co. 2001:677-89.
14. Cruz MJ, Kerschner JE, Beste DJ, Conley SF. Pierre Robin sequence: secondary respiratory difficulties and intrinsic feeding abnormalities, Laryngoscope 1999;109:1632-6.
15. Shprintzen RJ. The implications of the diagnosis of Robin Sequence. Cleft Palate Cran J 1992;29:205-9.
16. Shprintzen RJ. Pierre Robin, micrognahia, and airway obstruction: the dependency of treatment on accurate diagnosis. Int Anesth Clin 1988;26:64-71.
17. Sher AE. Mechanisms of airway obstruction in Robin Sequence: implication for treatment. Cleft Palate Cran J 1992;29:224-31.
18. Cohen MM. Editorial comment, Robin Sequences and complexes: Causal Heterogeneity and pathogenetic/phenotypic variability. Am J Med Genet 1999;84:311-5.
19. Olson TS, Kearns DB, Pransky SM, Seid AB. Early home management of patients with Pierre Robin sequence. Int J Pediatr Otorhi 1990;20:45-9.
20. Myer III CM, Reed JM, Cotton RT, Willhing JP, Shott SR. Airway management in Pierre Robin sequence. Otolaryngol Head Neck Surg 1998;118:630-5.
21. McCarthy JG, Schreiber J, Karp N. Lengthening of the human mandible by gradual distraction. Plast Reconstr Surg 1992;89:1-8.
22. Schaefer RD, Stadler, JAIII, Gosain AK. To distract or not to distract: An algorithm for airway management in isolated Pierre robin sequence. Plast Reconstr Surg 2004; 113:1113-1125.
23. Sher AE, Sphrintzen RJ, Thorphy MJ. Endoscopic observation of obstructive sleep apnea in children with anomalous upper airways: predictive and therapeutic value. Int J Pediatr Otorhi 1986;11:135-46.

以舌唇黏合術治療有呼吸道阻塞的 Pierre Robin Sequence病患：技術與結果

黃慧芬 羅綸洲[1] 陳昱瑞[1] 楊家森 牛震廣[2] 鍾美勇[2]

背 景： 呼吸道阻塞，餵食困難，可發生於 Pierre Robin sequence 的病患。在這一群病人中有些病人必須以外科方法才能改善呼吸道阻塞。手術適應症和方法及何時開刀乃有很多爭議。這個研究目的在於評估我們使用舌唇黏合術來處理因Pierre Robin sequence引起的上呼吸道阻塞的結果。

方 法： 從1995年3月到2002年5月，新生兒加護病房共有14位 Pierre Robin sequence 病患，因長期插管，體重增加遲緩或重覆呼吸道感染而接受舌唇黏合術。開刀方法是分別於舌頭及下唇各取一黏膜瓣後，將下唇及舌頭肌肉對縫。從舌及下唇取的黏膜則用來覆蓋在肌肉，並留置縫合線。我們評估病患術後的臨床反應。

結 果： 以舌唇黏合術治療的成功率是70%。14位病患中的10位有臨床改善，包括拔除插管，體重增加，回到居家照護，降低呼吸道感染，血氧濃度增加。舌唇黏合術是個簡單的步驟且沒有大的併發症。

結 論： 這個回溯研究顯示，有術前完整的呼吸道評估，舌唇黏合術可以成功治療部分嚴重上呼吸道阻塞的 Pierre Robin sequence 病患。當以外科治療呼吸道阻塞是必須時，且阻塞是因舌根部引起者，應優先考慮以舌唇黏合術治療。

(長庚醫誌2005;28:90-6)

關鍵字: 舌唇黏合術，呼吸道阻塞，Pierre Robin sequence 。

長庚紀念醫院 高雄院區 整形外科，[1]台北院區 顱顏中心，[2]高雄院區 小兒科
受文日期：民國93年10月27日；接受刊載：民國93年12月21日。
索取抽印本處：黃慧芬醫師，長庚紀念醫院 整形外科。高雄縣833鳥松鄉大埤路123號。Tel.: (07) 7317123轉8002; Fax: (07) 731- 7123轉8004; E-mail: spencerfaye@cgmh.org.tw