

PSEUDO-DUANE'S RETRACTION SYNDROME*

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(BY INVITATION), AND (BY INVITATION)

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IN 1905 ALEXANDER DUANE PUBLISHED A PAPER¹ DESCRIBING WHAT HAS COME TO BE known as Duane's retraction syndrome. Previous authors, particularly Silling (1887) and Turk (1896)² had written about the condition but Duane's description of 54 patients with his emphasis on retraction during attempted adduction has justifiably memorialized his name with this syndrome. Later investigators throughout subsequent decades have added to our basic concepts of this complex group of signs and symptoms.³ The etiology and pathogenesis is now, if anything, more complex than in those complacent days of yore. Nevertheless, recognition, if not always successful treatment, of Duane's syndrome has become almost a routine clinical exercise. It is said to comprise about 1% of all ocular muscle deviations. Our purpose is to call attention to a similar yet different set of symptoms and signs usually stemming from known etiology and to call the condition "pseudo-Duane's retraction syndrome."

CASE REPORTS

CASE 1

This 12-year-old black boy came to the emergency room complaining of diplopia on right gaze. His vision was 20/20 in each eye but he was unable to abduct his right eye 15° beyond the midline. The medical history and other ocular findings and physical signs were noncontributory. A diagnosis of sixth nerve palsy was considered and he was referred to the neuro-ophthalmology service. There he was noted to have a slight retraction (enophthalmus) upon attempted abduction. (Fig. 1). After further questioning he admitted falling on his face from an apple tree two days previously. Polytomograms showed clouding of the right ethmoid

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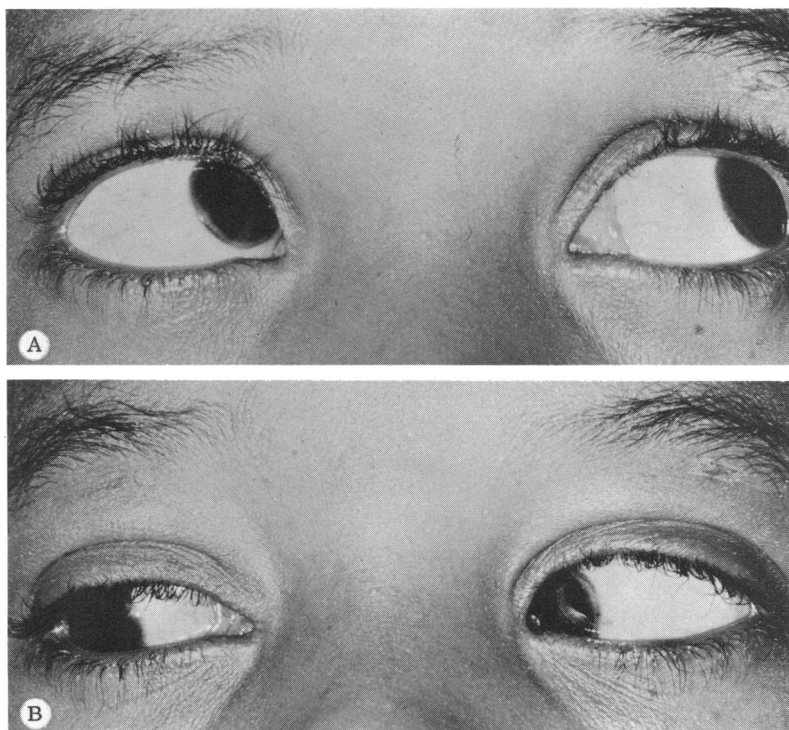


FIGURE 1

Retraction right eye on abduction A: Adduction (left gaze) normal B: Abduction (right gaze), restricted medial rectus with retraction and pseudoptosis.

sinus (Fig. 2). Forced ductions at the time of operation confirmed entrapment of the medial rectus of the right eye which was freed. There was less diplopia and greater abduction in the immediate postoperative period.

Comment: Usually in pseudo-Duane's retraction syndrome a history of trauma provides a major clue leading to the diagnosis. In this instance the patient's denial led the initial examiners astray. Retraction with an inability to abduct (pseudo-sixth nerve palsy) was the cardinal sign which ultimately led to a more accurate history as well as to a correct and treatable diagnosis.

CASE 2

A 12-year-old black boy stooped while playing volleyball and was struck in the right side of the face with a friend's knee. Fainting, a nose bleed, and limitation of



FIGURE 2

Polytomogram of Case 1. Cloudy ethmoidal sinus with fracture of the medial wall.

ocular motion warranted immediate admission to a general hospital where skull roentgenograms were read as normal. Six months later he was seen in consultation and the right eye was found to have limitation of adduction, and greater limitation of abduction with retraction occurring on the attempt. Vision was normal in each eye. Polytomograms showed a fracture of the ethmoids on the right side. Attempted forced abduction was definitely limited. A large anterior fracture just behind the medial canthal ligament and extending posteriorly was found at operation. Following repair the ductions improved. Diplopia remained in extreme right and left gaze.

Comment: The surgeon cannot depend upon the signs and symptoms nor the polytomograms to determine the extent of ethmoidal fracture and the degree of medial rectus entrapment. Rumelt and Ernest described a patient who showed no retraction with narrowing of the palpebral fissure "presumably because the entrapment was so far posterior in the orbit."⁴ The skin-orbital approach with wide exposure is the preferred operative procedure.

CASE 3

A 34-year-old white woman with a questionable previous diagnosis of multiple sclerosis fell in her bathroom and struck her left orbital area on a sink. There was spontaneous bleeding from the nose which ceased without treatment. She noticed diplopia on left gaze almost immediately. Examination two days later showed a restriction of abduction with concurrent retraction of the left eye, and she complained of diplopia on left lateral gaze. There were periorbital and subconjunctival ecchymoses and edema on the left side. Intranasal examination revealed medial displacement of the middle turbinate on the left side. Polytomograms demonstrated blurring over the posterior ethmoid air cells with medial displacement of the lamina papyracea. Entrapment was confirmed with forced duction tests and was relieved with medial wall surgery. Postoperative diplopia and limitation of abduction were present only in extreme lateral gaze.

Comment: Nasal bleeding and intranasal deformity may serve as clues suggesting combined orbital and nasal trauma incriminating the ethmoidal cells of the medial orbital wall.⁵

Wide exposure and careful inspection revealed a more extensive fracture than was anticipated from preoperative study. This would seem to condemn the transconjunctival approach which has its advocates.⁶

CASE 4

Courtesy of Dr Joseph C. Flanagan. A 25-year-old white man was hit with a sledge hammer in the region of the right orbit one and one half years previously. He had four surgical procedures performed earlier including repair of the orbital floor. Vision in the right eye was 20/80 (central retinal pigmentary changes and mild optic atrophy were present); vision in the left eye was 20/20. The right orbit was deformed. Abduction was almost absent and elevation and depression were greatly restricted. On right gaze enophthalmic retraction was present and the patient noted diplopia. The left eye was normal. Polytomography revealed, among other orbital and facial deformities, a fracture of the ethmoidal wall. Surgical repair was difficult. Monitoring effectiveness of release of the medial rectus by attainable degree of forced duction finally culminated in satisfactory results.

Comment: Medial wall blowout fracture may (and often does) accompany orbital floor blowout fractures,^{7,8} this being an extreme example.

CASE 5

A 60-year-old white woman came to the clinic because of inability to abduct her left eye and because of retraction noted upon the attempt. Her ocular history was noncontributory. She was thought to have a sixth nerve palsy. Some limitation of adduction was also noted. Roentgenograms revealed invasion of the orbital walls (Fig. 3) especially the medial wall with probable medial rectus en-

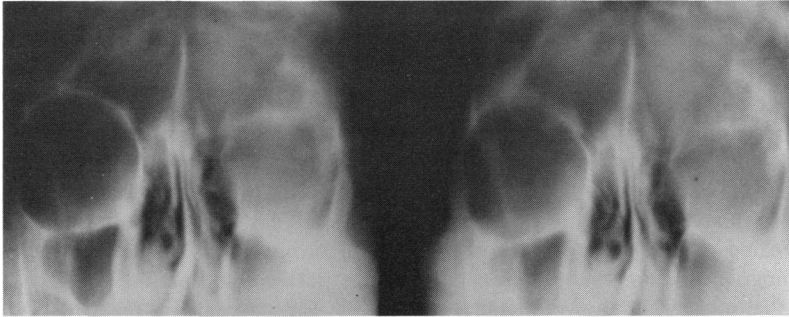


FIGURE 3

Polytomograms of Case 5. Clouding of the left orbit due to invasion by what later proved to be metastatic breast cancer.

trapping. The forced duction test was positive. Further medial consultation uncovered a primary carcinoma of the left breast with left orbital metastases.

Comment: Again history is of importance. If a history of trauma is missing, then some other cause of limitation and retraction during attempted abduction must be sought. Roentgenograms and forced duction tests are indicated in all instances of unexplained retraction.

DISCUSSION

It is not the purpose of this report to delve into ramifications of the well known Duane's syndrome. This has been adequately reviewed in recent texts.^{3,9} The goal is to highlight the differences between the standard form of Duane's syndrome and pseudo-Duane's syndrome.

In the first place, the classic Duane's syndrome primarily affects the lateral rectus which usually because of its fibrotic condition resists adduction and accounts for absence of abduction. With medial rectus entrapment just the opposite is present—namely inability to abduct because of involvement of the medial rectus. In both instances (that is with adduction in the former and abduction in the latter) a narrowing of the palpebral fissure appears with a retraction enophthalmus and pseudoptosis occurring secondary to the attempted movement. All horizontal gaze movements may be limited in both types. Forced duction tests are consistent with these findings and indeed help in a major way to elucidate the extent of muscular involvement. However, the etiology and pathogenesis of the two syndromes are entirely different. Duane's syndrome is thought to be due to a fibrosis of the lateral rectus with or without similar changes in the medial rectus and with possible central nervous

system (brain stem) disease leading to a cocontracture of the recti muscles. Orbital roentgenograms are of little value in this condition. Pseudo-Duane's retraction syndrome is most often due to trauma of the medial wall ethmoidal sinuses with entrapment of the medial rectus extending through the fractured lamina papyracea. It may occur secondary to other changes in the orbit (see Case 5). We are now collecting a series of thyroid myopathy patients who demonstrate retraction with attempted abduction. As is true for orbital trauma in general, roentgenograms, particularly polytomograms, are essential in making or confirming the diagnosis. Differences between the classic Duane's syndrome and pseudo-Duane's syndrome are listed in Table I.

There are thus some cardinal features of the pseudo-Duane's retraction syndrome which should be highlighted. The review of our patients and a search of the literature indicates that age, sex, color, and side of involvement have no significance.^{9,10,11} Roentgenograms in the form of polytomograms (and probably computerized axial tomography when it becomes more generally available) are of maximum help and are essential to the diagnosis when medial rectus entrapment is suspected.^{5,11-19} Forced duction (traction) tests,²⁰ best performed at the time of surgery, not only help establish the degree of entrapment of the medial rectus but also serve as indicators as to whether the malady has been alleviated with attempted surgical intervention. Witness the report of the surgeon treating Case 4: "A lid speculum was placed and forced duction tests were performed on the right eye and there was marked limitation of abduction of the right eye. An incision was made along the superior nasal rim of the orbit below the area of the trochlea. The periosteum was elevated to expose the orbital rim and also was elevated at the upper border of the lacrimal fossa and below the trochlea. A fracture line was

TABLE I: THE RETRACTION SYNDROME

	Classic Duane's Syndrome	Pseudo-Duane's Syndrome
History	Present since early age; no diplopia; frequently amblyopia	Trauma; diplopia common
Ductions:		
Abduction	Absent usually	Absent usually; narrowing of fissure; RETRACTION on attempt
Adduction	Restricted; tendency to up or down shoot; narrowing of fissure; RETRACTION on attempt	Absent or present
Polytomograms	Normal	Fracture; frequently air in orbit
Surgical Prognosis	Fair to poor	Excellent

 TABLE II: CARDINAL FEATURES OF PSEUDO-DUANE'S RETRACTION SYNDROME

History: trauma, highly significant
Abduction: absent; retraction on attempt
Polytomes: highly contributory
Forced Duction: positive for restriction of medial rectus
Surgical Prognosis: excellent, adequate exposure necessary

seen at the orbital rim. It extended back into the ethmoid air cells. There was tissue prolapsed into the ethmoid air cells and this was elevated with the periosteal elevator and replaced in the orbit. Forced duction tests were again performed and there was moderate alleviation of the restriction; however, some restriction remained and therefore more tissue was freed from the ethmoid bone with the periosteal elevator. Forced ductions were again attempted and the eye could be abducted fully. The periosteum was closed with #4-0 chromic catgut and the skin was closed with interrupted #6-0 black silk and a pressure dressing applied."

Surgical prognosis ranges from fair to excellent in pseudo-Duane's retraction syndrome except in such entities as metastatic orbital disease (Case 5). In general, the longer the condition has been present the poorer will be the prognosis for full recovery of lateral motion with complete elimination of lateral gaze diplopia. Unlike the classic Duane's retraction syndrome, amblyopia is no problem in these patients. In our opinion wide surgical exposure of the medial wall is mandatory. Frequently the fracture extends farther posteriorly than was suspected. Again, forced duction tests should be performed before closing the incision (Table II).

We agree with Parks⁹ and find classification of Duane's retraction syndrome in the various types, classes, and varieties²¹⁻²³ to be more confusing than enlightening. Consequently we have concluded that the terminology most appropriate for the entrapped medial rectus to be: "pseudo-Duane's retraction syndrome" and have carefully avoided such words as "reverse", "inverse", and "mirror image" because we found them either inaccurate or conflicting with our observations.

SUMMARY

Five patients presented with signs that were similar to but opposite from Duane's retraction syndrome. Most had a history of orbital trauma. On attempted abduction a narrowing of the palpebral fissure and retraction of the globe was observed. Diplopia with lateral gaze was present. Roentgenograms (polytomograms) showed involvement of the medial orbital wall. Forced duction tests were positive. Surgical repair of the fracture

and release of the entrapped muscle as determined by forced duction tests and by postoperative motility led to successful results.

REFERENCES

1. Duane A: Congenital Deficiency of Abduction, Associated with Impairment of Adduction, Retraction Movements. Contractions of the Palpebral Fissure and Oblique Movements of the Eye. *Arch Ophthalmol* 34:133-159, 1905.
2. Duke-Elder S (ed): Ocular Motility and Strabismus, *System of Ophthalmology*. St. Louis, CV Mosby, 1973, vol 6, pp 740-746.
3. Burian HM, von Noorden GK: *Binocular Vision and Ocular Motility; Theory and Management of Strabismus*. St. Louis, CV Mosby, 1974, pp 368-391.
4. Rumelt MB, Ernest JT: Isolated Blowout Fracture of the Medial Orbital Wall with Medial Rectus Muscle Entrapment. *Am J Ophthalmol* 73:451-453, 1972.
5. Valvassori GE, Hord GE: Traumatic Sinus Disease. *Seminars in Roentgenology* 3:160-171, 1968.
6. Schonder AA, Salz JJ, Magnus WW, Castner DV: A Conjunctival Approach to Repair of Fracture of Medial Wall of Orbit. *Ann Ophthalmol* 4:297-308, 1972.
7. Duke-Elder S (ed): Mechanical Injuries, *System of Ophthalmology*. St. Louis, CV Mosby, 1972, vol 14, part 1, pp 248-249.
8. Dodick JM, Galin MA, Littleton JT, Sod LM: Concomitant Medial Wall Fracture and Blowout Fracture of the Orbit. *Arch Ophthalmol* 85:273-276, 1971.
9. Parks MM: *Ocular Motility and Strabismus*. Hagerstown, Harper & Row, 1975, pp 165-167.
10. Miller GR, Glaser JS: The Retraction Syndrome and Trauma. *Arch Ophthalmol* 76:662-663, 1966.
11. Edwards WC, Ridley RW: Blowout Fracture of Medial Orbital Wall. *Am J Ophthalmol* 62:248-249, 1968.
12. Fischbein FI, Lesko WS: Blowout Fracture of the Medial Orbital Wall. *Arch Ophthalmol* 81:162-163, 1969.
13. Converse JM: Two Plastic Operations for Repair of Orbit Following Severe Trauma and Extensive Comminuted Fracture. *Arch Ophthalmol* 31:323-325, 1944.
14. Smith B, Regan WF: Blowout Fracture of the Orbit. *Am J Ophthalmol* 44: 733-739, 1957.
15. Converse JM, Smith B: Naso-Orbital Fractures. *Trans Am Acad Ophthalmol Otolaryngol* 67:622-634, 1963.
16. Jones IS: X-Ray Findings. *Trans Am Acad Ophthalmol Otolaryngol* 67:635-642, 1963.
17. Fueger GF, Milauskas AT, and Britton W: The Roentgenologic Evaluation of Orbital Blow-out Injuries. *Am J Roentgenol Radium Ther Nucl Med* 97:614-617, 1966.
18. Gould HR, Titus CO: Internal Orbital Fractures: The Value of Laminography in Diagnosis. *Am J Roentgenol Radium Ther Nucl Med* 97: 618-623, 1966.
19. Trokel ST, Potter GD: Radiographic Diagnosis of Fracture of the Medial Wall of the Orbit. *Am J Ophthalmol* 67:772-773, 1969.
20. Duke-Elder S (ed): Mechanical Injuries, *System of Ophthalmology*. St. Louis, CV Mosby, 1972, vol 14, part 1, pp 254-255.
21. Malbran E: *Estrabismos y Paralisis*, Buenos Aires, 1949, pp 627-637.
22. Brown HW, Allen JH (ed): *Strabismus Ophthalmic Symposium*, St. Louis, CV Mosby, 1950, p 205.
23. Huber A: Duane's Retraction Syndrome: Considerations on Pathogenesis and Etiology of the Different Forms of Duane's Retraction Syndrome. In *Strabismus*, St. Louis, CV Mosby, 1969, p 26.

DISCUSSION

DR PHILIP KNAPP. I would like to thank the authors for sending me this paper in January and congratulate Doctor Duane, Doctor Schatz, and Doctor Caputo for

finding and presenting these five cases. I certainly think it's time that someone named "Duane" finally got back into the motility field.

The authors have described five cases of limited abduction with narrowing of the lid fissure on attempted abduction. Four of those followed trauma and one patient had metastatic cancer. They stress that refined roentgenographic technique such as hypocycloidal tomography and computerized tomography are helpful in making the diagnosis. However, when you get in the operating room, the traction test will be positive and this is most helpful. The authors like a cutaneous approach. They operate between the trochlea and the lacrimal region nasally. I don't think this is necessary. We have only had three cases which just goes to prove that there is more trauma in Philadelphia than in New York. Anyway, I think you can probably use the conjunctival approach. You can open the conjunctiva widely with the lid speculum removed and get good exposure. If you don't, you can always go in with the lower lid incision used in floor fractures. Many of these cases also have a floor fracture. The commonest type is the combined floor and the nasal wall fracture. I think it makes it much easier to do your traction test if you have the conjunctiva open and the medial rectus exposed with either a suture or a muscle hook under it. In that way you can tell when the muscle is completely free.

The authors call these cases the atypical Duane's retraction syndrome. The typical syndrome includes the following: there is no abduction, there is widening when the patient tries to abduct, it is usually the left eye, it is more common in females, and the adduction is also poor. This, of course, is what differentiates this disorder from a VI nerve paresis. What gives it its name is that when the patient tries to adduct the eye, retraction of the globe produces a narrowing of the lid fissure and you may get either an up or downshoot. This picture was originally blamed on a fibrosis of the lateral rectus. It doesn't adduct and the medial rectus pulling against this fibrous muscle caused the eye to pull back and that gave the retraction. Electromyography certainly shows that the retraction is due to paradoxical or some type of aberrant innervation. In other words, when the lateral rectus is supposed to abduct it is silent, but when it adducts and is supposed to be silent, it fires. This co-contraction is what gives the retraction. I think this is the most likely etiology. I have looked at a lot of lateral recti in Duane's because I have frequently recessed it on the three recession procedure and they have all looked normal. I have never seen a fibrous lateral rectus as originally described. Another thing is that practically any mechanical basis of squint will give you a limited duction. For instance, you do a large recession-resection and afterwards it doesn't move. For example, after a recession-resection for esotropia the eye doesn't adduct. If the fissure widens on attempted adduction it usually means a slipped medial rectus. If it is due to excessive resection or fibrosis around the lateral rectus, there is usually narrowing on attempted adduction. In other words, these cases are really just another form of mechanical squint and I wonder if we need to call it a syndrome.

In conclusion, I'd like to compliment the authors for presenting these cases and getting people to think mechanical.

DR GUNTER K. VON NOORDEN. The choice of the term pseudo-Duane's syndrome is not a fortunate one in describing this condition since retraction occurs on attempted abduction and not in adduction as in true Duane's syndrome. It would be preferable to speak of a "reverse" Duane's syndrome.

Not all of these are caused by fracture of the medial orbital wall as there may be other mechanical factors responsible for causing retraction on attempted abduction. I recently saw a patient who had several operations for a pterygium involving the nasal aspects of the conjunctiva and cornea. Conjunctival scarring was so extensive that abduction was severely limited and attempts to abduct the globe caused retraction.

DR RICHARD D. RICHARDS. We had two of these cases, and we approached these from the conjunctival approach and followed it by placing the muscle hook under the medial rectus and then passing the other muscle hook between the medial rectus and the globe. We were able to feel the point of obstruction where the muscle was caught and could free it quite easily this way and didn't find it was necessary to do an orbital approach. We didn't find that we could actually see the area that was caught but by manipulating one muscle hook against the other and pulling on one and pushing the other back and forth you could feel the area and free it. We found this quite effective.

DR THOMAS DUANE. I thank Doctor Knapp for his kind remarks. We seem to have a difference of opinion in Philadelphia and New York. Of course, as he says we're the city of brotherly love and we probably see more trauma than they do. He took issue with several things, but first of all he didn't like the use of the type of exposure that we recommend. The first to recommend conjunctival exposure and popularize it were Galin and his associates from New York. The group from Chicago, Ernest and his co-workers, found in one patient that the conjunctival approach failed because the entrapment was so far back in the orbit that they simply couldn't reach back there with the conjunctival approach. In fact, they had to back out and redo it; and so they were the first to stress the approach through the skin. It's not a matter of whether you like one or the other. If you can free it that's the important thing.

I would like to discuss a point that Doctor von Noorden brought up. There seems to be a lot of quibbling about whether this is the right name. Whether it's a syndrome and whether it should be called "reverse." I originally called it the "reverse Duane syndrome." We studied a patient who had carcinoma rather than trauma. We found that in 1949, Malbran had described in a book, *Strabismus with Paralysis*, three different types of Duane's syndrome. I had Juan Arentsen translate this for me and it became so confused I couldn't make any sense out of it even though it was translated into good English. In 1950, the next year, Dr Harold W. Brown described, in a symposium in New Orleans, a whole group of Duane's. He has talked to me personally and said he's never seen one that didn't have limitation of adduction. He thinks I've chosen the wrong name also. In 1969 Doctor Huber described three types of Duane's syndromes. Hence it's been called reverse Duane, it's been called inverse Duane, and

it's been called mirror image Duane. The big thing about the name is not only whether it fits or whether it denotes the salient change or trouble with abduction as opposed to trouble with adduction but the reason we call it "pseudo-Duane" is because the etiology is completely different. Co-contraction and the paradoxical innervation with fibrosis is an entirely different problem from that which is due to entrapment of the medial rectus. For this reason, even though there is a lot of confusion in forms of Duane's syndromes we still feel "pseudo-Duane" is the most appropriate title.

Thank you.

DR THOMAS DUANE. (The following day). I'm trying to collect a series of patients that have retraction involving the medial rectus with a nontraumatic origin. Those of you who have case histories of any such patients I would like to have you notify me if you would—send the case reports to me and I would appreciate it very much.

In addition, I want to tell you about the problem I have. I put in a bad night last night. Too many people came around to me at a party in the North Garden and asked why I persisted in calling the paper that I gave yesterday "pseudo-Duane" when other titles seem more appropriate. I thought I had given a scientific explanation of this but evidently a number of people weren't convinced and so later in the evening in my heart of hearts I really started wondering why I did call it that and I would like to give a little background.

In the first place I'm not related at all to Alexander Duane. My grandfather came over from Ireland right after the Civil War and ended up in the Midwest. We've certainly never been confused with aristocracy whereas Alexander Duane was part of the New York aristocracy. I think he was related to Ben Franklin and other people who lived in that era by marriage and so forth. It's kind of quixotic that Philip Knapp was chosen to discuss my paper because in 1948 I rode on the subway with Arnold Knapp, his father, from 165th Street to 42nd Street. For those of you who don't know, he was the Dean of American Ophthalmology. I asked him who was the greatest all around ophthalmologist he had ever known and he told me without a moment's hesitation that it had to be Alexander Duane.

[Slide] Alexander Duane was a very distinguished ophthalmologist, the soul of propriety, who was once the President of this august Society. Last night about midnight my room lit up. It didn't awaken Julia, my wife, but it did me and I looked up and there was Alexander Duane.

He said: "That wasn't much of a paper you gave. It wasn't an original observation in the first place."

I said: "Neither was yours."

"But," he answered, "at least I reported 54 cases and you had only five and as you said you stumbled on the first one, you even made the wrong diagnosis."

I said: "Well, okay."

"Furthermore," he said "you know you've been drinking it up, eating it up, and have gone halfway around the world to present a paper while wearing modified pajamas and tennis shoes. And," he said, "you called your oldest daughter Alexa so I really think you're trying to capitalize on me. Indeed as I think it over you must be the biggest pseudo-Duane in the history of medicine."